Table of Contents

As filed with the Securities and Exchange Commission on May 14, 2018.

Registration No. 333-224488

UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

Amendment No. 1
to

FORM S-1

REGISTRATION STATEMENT
UNDER
THE SECURITIES ACT OF 1933

Kiniksa Pharmaceuticals, Ltd.

(Exact name of registrant as specified in its charter)

Bermuda	**2834**	**98-1327726**
(State or other jurisdiction of incorporation or organization)	(Primary Standard Industrial Classification Code Number)	(I.R.S. Employer Identification No.)

Clarendon House
2 Church Street
Hamilton HM11, Bermuda
+1 (441) 295-5950

(Address, including zip code, and telephone number, including area code, of registrant's principal executive offices)

Kiniksa Pharmaceuticals Corp.
100 Hayden Avenue
Lexington, MA 02421
(781) 431-9100

(Name, address, including zip code, and telephone number, including area code, of agent for service)

Copies to:

Johan V. Brigham **Nathan Ajiashvili** **Stephen W. Ranere** **Latham & Watkins LLP** **200 Clarendon Street,** **27th Floor** **Boston, Massachusetts 02116** **(617) 948-6000**	**Alan Dickson** **Chiara T. Nannini** **Conyers Dill & Pearman Limited** **Clarendon House,** **2 Church Street** **PO Box HM 666** **Hamilton, HM CX, Bermuda** **+1 (441) 295-1422**	**Patrick O'Brien** **Michael S. Pilo** **Ropes & Gray LLP** **Prudential Tower** **800 Boylston Street** **Boston, Massachusetts 02199** **(617) 951-7000**

Approximate date of commencement of proposed sale to the public:
As soon as practicable after this Registration Statement is declared effective.

If any of the securities being registered on this form are to be offered on a delayed or continuous basis pursuant to Rule 415 under the Securities Act of 1933, check the following box. o

If this form is filed to register additional securities for an offering pursuant to Rule 462(b) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. o

If this form is a post-effective amendment filed pursuant to Rule 462(c) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. o

If this form is a post-effective amendment filed pursuant to Rule 462(d) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. o

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, or a smaller reporting company. See the definitions of "large accelerated filer," "accelerated filer" and "smaller reporting company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer o	Accelerated filer o	Non-accelerated filer ☒ (Do not check if a smaller reporting company)	Smaller reporting company o Emerging growth company ☒

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 7(a)(2)(B) of the Securities Act. ☒

CALCULATION OF REGISTRATION FEE

Title of Each Class of Securities To Be Registered	**Amount to be Registered(1)**	**Proposed Maximum Offering Price Per Share(2)**	**Proposed Maximum Aggregate Offering Price(3)**	**Amount of Registration Fee(3)**
Class A Common Shares, $0.000273235 par value per share	8,050,000	$19.00	$152,950,000	$19,043

(1) Includes 1,050,000 shares that the underwriters have the option to purchase.

(2) Estimated solely for the purpose of calculating the registration fee pursuant to Rule 457(a) under the Securities Act of 1933, as amended.

(3) $12,450 of this registration fee was previously paid by the Registrant in connection with the filing of its Registration Statement on Form S-1 on April 27, 2018.

The Registrant hereby amends this Registration Statement on such date or dates as may be necessary to delay its effective date until the Registrant shall file a further amendment which specifically states that this Registration Statement shall thereafter become effective in accordance with Section 8(a) of the Securities Act of 1933 or until the Registration Statement shall become effective on such date as the Commission, acting pursuant to said Section 8(a), may determine.

Table of Contents

The information in this prospectus is not complete and may be changed. We may not sell these securities until the registration statement filed with the Securities and Exchange Commission is effective. This prospectus is not an offer to sell these securities and it is not soliciting an offer to buy these securities in any jurisdiction. where the offer or sale is not permitted

Subject to Completion, Dated May 14, 2018

PRELIMINARY PROSPECTUS

7,000,000 Shares

Class A Common Shares

This is an initial public offering of our Class A common shares. All 7,000,000 Class A common shares are being sold by us.

Prior to this offering, there has been no public market for our Class A common shares. It is currently estimated that the initial public offering price per share will be between $17.00 and $19.00. We have applied to have our Class A common shares listed on The Nasdaq Global Market under the symbol "KNSA."

We are an "emerging growth company" as defined in Section 2(a) of the Securities Act of 1933, as amended, as modified by the Jumpstart Our Business Startups Act of 2012, and as such have elected to comply with certain reduced public company reporting requirements for this prospectus and future filings. See "Prospectus Summary — Implications of Being an Emerging Growth Company."

Investing in our Class A common shares involves risk. See "Risk Factors" beginning on page 13 to read about factors you should consider before buying our Class A common shares.

Neither the Securities and Exchange Commission nor any other regulatory body has approved or disapproved of these securities or passed upon the accuracy or adequacy of this prospectus. Any representation to the contrary is a criminal offense.

	Per Share	Total
Initial public offering price	$	$
Underwriting discounts and commissions[1]	$	$
Proceeds, before expenses, to Kiniksa Pharmaceuticals, Ltd.	$	$

(1) See "Underwriting" beginning on page 208 for additional information regarding underwriting compensation.

Following this offering, we will have four classes of common shares outstanding: Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares. All classes of our common shares will be economically equivalent to each other. The rights of the holders of our Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares will be identical, except with respect to voting, conversion and transferability. Each Class A common share will be entitled to one vote and will not be convertible into any other class of our share capital. Each Class B common share will be entitled to ten votes and will be convertible at any time at the election of the holder into one Class A common share or one Class B1 common share and will automatically convert into Class A common shares upon transfer to an unaffiliated party. The rights of the holders of our Class A1 common shares and Class B1 common shares will be identical, except with respect to conversion. Each Class A1 common share and Class B1 common share will have no associated voting rights. Each Class A1 common share will be convertible into one Class A common share, subject to certain limitations, as described in this prospectus. Each Class B1 common share will be convertible into one Class A common share or one Class B common share, subject to certain limitations, as described in this prospectus. Immediately following this offering, the holders of Class A common shares will account for 22.2% of our aggregate voting power and the holders of Class B common shares will account for the remaining 77.8% of our aggregate voting power. See "Description of Share Capital — Common Shares" for more information on the rights of the holders of our Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares.

We have granted the underwriters the option to purchase up to an additional 1,050,000 of our Class A common shares for a period of 30 days after the date of this prospectus.

Certain of our existing shareholders, including shareholders affiliated with one of our directors, have indicated an interest in purchasing an aggregate of approximately $50.0 million of Class A common shares in this offering at the initial public offering price per share. However, because indications of interest are not binding agreements or commitments to purchase, the underwriters may determine to sell more, less or no shares in this offering to any of these existing shareholders, or any of these existing shareholders may determine to purchase more, less or no shares in this offering. The underwriters will receive the same underwriting discount on any shares purchased by these existing shareholders as they will on any other shares sold to the public in this offering.

The underwriters expect to deliver the Class A common shares to investors against payment on or about , 2018.

Goldman Sachs & Co. LLC **J.P. Morgan**

JMP Securities **Wedbush PacGrow**

Prospectus dated , 2018

TABLE OF CONTENTS

We have not, and the underwriters have not, authorized anyone to provide any information or to make any representations other than those contained in this prospectus or in any free writing prospectus prepared by or on behalf of us or to which we have referred you. We take no responsibility for, and can provide no assurance as to the reliability of, any other information that others may give you. This prospectus is an offer to sell only the shares offered hereby, and only under circumstances and in jurisdictions where it is lawful to do so. The information contained in this prospectus or in any applicable free writing prospectus is current only as of its date, regardless of its time of delivery or any sale of our Class A common shares. Our business, financial condition, results of operations and prospects may have changed since that date.

For investors outside the United States: We have not, and the underwriters have not, done anything that would permit this offering or possession or distribution of this prospectus in any jurisdiction where action for that purpose is required, other than in the United States. Persons outside the United States who come into possession of this prospectus must inform themselves about, and observe any restrictions relating to, the offering of the Class A common shares and the distribution of this prospectus outside the United States.

TRADEMARKS

We own or have rights to trademarks that we use in connection with the operation of our business, including Kiniksa™ and ARCALYST®. Kiniksa™ is a trademark of Kiniksa Pharmaceuticals, Ltd. and ARCALYST® is a trademark of Regeneron Pharmaceuticals, Inc. Solely for convenience, trademarks, service marks and trade names referred to in this prospectus, including Kiniksa and ARCALYST, are listed without the ®, SM and ™ symbols. We will assert, to the fullest extent under applicable law, our rights to our intellectual property. Trademarks, service marks and trade names of third parties are the intellectual property of such parties.

PROSPECTUS SUMMARY

This summary highlights information contained elsewhere in this prospectus. This summary does not contain all of the information you should consider before investing in our Class A common shares. You should read this entire prospectus carefully, especially the sections entitled "Risk Factors" and "Management's Discussion and Analysis of Financial Condition and Results of Operations" and our consolidated financial statements and the related notes appearing at the end of this prospectus, before making an investment decision. This prospectus includes forward-looking statements that involve risks and uncertainties. See "Special Note Regarding Forward-Looking Statements."

As used in this prospectus, unless the context otherwise requires, references to "we," "us," "our," the "Company" and "Kiniksa" refer to Kiniksa Pharmaceuticals, Ltd. and its consolidated subsidiary, together.

Overview

We are a clinical-stage biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have a pipeline of product candidates across various stages of development, currently focused on autoinflammatory and autoimmune conditions. We have three clinical-stage product candidates, one of which is anticipated to commence a Phase 3 clinical trial in 2018. We follow a disciplined and methodical approach to selectively identify and acquire product candidates with strong biologic rationales or validated mechanisms of action. We believe that each of our product candidates has the potential to address multiple indications.

Our Programs

- **Rilonacept** (ARCALYST) is a protein for inhibiting interleukin-1a and interleukin-1b. Cytokines are small proteins that play a key role in cell signaling. Rilonacept is approved by the U.S. Food and Drug Administration, or FDA, for the treatment of cryopyrin-associated periodic syndromes, or CAPS, and has been commercially available from Regeneron Pharmaceuticals, Inc., or Regeneron, for this indication since 2008. We are initially developing rilonacept for the treatment of recurrent pericarditis, a debilitating inflammatory cardiovascular disease. We are not aware of any therapy currently approved by the FDA for the treatment of recurrent pericarditis. We are currently conducting an open-label Phase 2 proof-of-concept clinical trial in this disease and expect to report preliminary data in 2018. The trial is divided into five parts to evaluate rilonacept in various subgroups of patients with pericarditis. As of April 23, 2018, a total of four subjects were enrolled in the clinical trial. Three of the four subjects were experiencing symptomatic recurrent pericarditis at the time of enrollment. All three subjects showed a reduction in C-reactive protein measurements, as well as a reduction in scores of an 11-point Numerical Rating Scale instrument for assessing pericardial-associated chest pain, in the first week of treatment. The fourth subject, in a separate patient cohort, was on corticosteroids at the time of enrollment but not experiencing active symptoms, and had a history of corticosteroid dependence. This subject was treated with rilonacept and as of April 23, 2018, had been free of pericarditis flares for three weeks. Among these subjects, the most commonly reported adverse reaction associated with rilonacept has been injection-site reactions.

- **Mavrilimumab** is a monoclonal antibody that antagonizes the signaling of granulocyte macrophage colony stimulating factor, or GM-CSF. We are focusing our initial development efforts for mavrilimumab on giant cell arteritis, or GCA, an inflammatory disease of the blood vessels with unmet medical need that can lead to blindness if left untreated. MedImmune

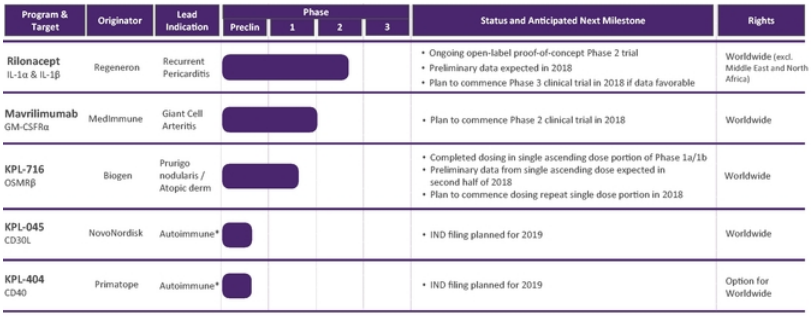
Limited, or MedImmune, initially developed mavrilimumab for the treatment of rheumatoid arthritis, or RA. MedImmune's Investigational New Drug application, or IND, for the clinical development of mavrilimumab for the treatment of RA was initially put on clinical hold in 2010 before human data had been generated due to certain effects that were observed in non-clinical studies, which coincides with a theoretical risk of developing pulmonary alveolar proteinosis, or PAP, possibly in the setting of GM-CSF inhibition. Since then, in 2014, the FDA acknowledged that clinical studies in refractory RA may be appropriate based on MedImmune's clinical studies in Europe in which it dosed over 550 RA patients with mavrilimumab with no evidence of PAP. MedImmune has since withdrawn the IND for mavrilimumab for the treatment of RA. We intend to develop mavrilimumab for the treatment of GCA under a new IND in the United States and new Clinical Trial Application, or CTA, in Europe, and plan to initiate a Phase 2 clinical trial in 2018.

- **KPL-716** is a monoclonal antibody that simultaneously inhibits the signaling of the cytokines interleukin-31, or IL-31, and oncostatin M, or OSM, by targeting their common receptor subunit, oncostatin M receptor beta, or OSMRb. We plan to study KPL-716 in a variety of pruritic and fibrotic indications driven by these cytokines and we believe KPL-716 is the only monoclonal antibody in development that simultaneously targets both pathways. We are currently enrolling subjects in a Phase 1a/1b clinical trial in healthy volunteers and in subjects with atopic dermatitis as a proof-of-concept for pruritic conditions. We have completed dosing in the first portion of the Phase 1a/1b clinical trial utilizing a single ascending dose design. Each dose cohort in the first portion received a single dose of KPL-716 administered either intravenously or subcutaneously. We expect to report preliminary data from the single ascending dose cohorts of this portion of the trial in the second half of 2018. In the second portion of the Phase 1a/1b clinical trial, each subject in the cohort will receive repeated single doses of KPL-716 administered subcutaneously. We plan to commence dosing in this portion of the trial in 2018, subject to submission to, and review by, the FDA and Canadian regulatory authorities of our recently completed primate chronic toxicology study. If the data from the Phase 1a/1b clinical trial are favorable, we expect our two initial targeted indications for future development of KPL-716 to be prurigo nodularis and atopic dermatitis, both inflammatory, pruritic skin conditions with unmet medical need.

- **KPL-045** is a monoclonal antibody inhibitor of the CD30/CD30L interaction, a T-cell co-stimulatory receptor involved in activated T-memory cell function. We are planning IND-enabling studies in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in 2019.

- **KPL-404** is a monoclonal antibody inhibitor of the CD40/CD40L interaction, a central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity. We are planning IND-enabling studies in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in 2019.

The following table summarizes our current pipeline of product candidates:



Program & Target	Originator	Lead Indication	Phase	Status and Anticipated Next Milestone	Rights
Rilonacept IL-1α & IL-1β	Regeneron	Recurrent Pericarditis	Preclin	• Ongoing open-label proof-of-concept Phase 2 trial • Preliminary data expected in 2018 • Plan to commence Phase 3 clinical trial in 2018 if data favorable	Worldwide (excl. Middle East and North Africa)
Mavrilimumab GM-CSFRα	MedImmune	Giant Cell Arteritis	Preclin	• Plan to commence Phase 2 clinical trial in 2018	Worldwide
KPL-716 OSMRβ	Biogen	Prurigo nodularis / Atopic derm	Preclin	• Completed dosing in single ascending dose portion of Phase 1a/1b • Preliminary data from single ascending dose expected in second half of 2018 • Plan to commence dosing repeat single dose portion in 2018	Worldwide
KPL-045 CD30L	NovoNordisk	Autoimmune*	Preclin	• IND filing planned for 2019	Worldwide
KPL-404 CD40	Primatope	Autoimmune*	Preclin	• IND filing planned for 2019	Option for Worldwide

Notes:
- Rilonacept (ARCALYST®) is approved and marketed for cryopyrinassociated periodic syndrome, in the United States by Regeneron. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication.

* We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.

In addition to the indications described above, we plan to evaluate rilonacept, mavrilimumab and KPL-716 in other indications. We also plan to be opportunistic in our business development activities to identify and potentially acquire the rights to additional programs. We have also initiated our own internal research efforts to discover and develop molecules to address areas of unmet medical need.

We intend to directly commercialize our product candidates, if approved, in the United States and select international markets. In parallel with our product development timelines, we plan to build our own commercial and operational organizations around the world. We anticipate building targeted medical affairs and sales teams focused on specialist physicians who treat the patient populations addressed by our product candidates.

Our Team

We have assembled an experienced management team with a successful track record, many of whom have previously worked together at companies that developed and commercialized therapeutics for underserved, rare and specialty-focused patient populations. Our team has expertise across the spectrum of global drug discovery, development, manufacturing and commercialization activities in diseases within both large and orphan indications. Our Chairman and Chief Executive Officer, Sanj K. Patel, has more than 25 years of scientific, clinical and commercial experience in the pharmaceutical and biotechnology industries. Our Chief Medical Officer, John F. Paolini, M.D., Ph.D., has more than 15 years of experience planning, operating and executing clinical development programs across a range of disease indications from orphan diseases to large cardiovascular diseases, and ten years as a practicing cardiologist.

Our Strategy

Our vision is to build a fully-integrated global biopharmaceutical company by discovering, acquiring, developing and commercializing life-changing therapies for debilitating diseases. We are currently developing a pipeline of novel drug product candidates for the treatment of autoinflammatory and autoimmune diseases, and we aim to be an industry leader in these areas.

We are pursuing multiple programs in parallel, with the goal of delivering safe and effective therapies to patients as efficiently as possible.

Critical components of our business strategy include the following:

- **Efficiently and rapidly advance our product candidates through the development process.** We believe that our product candidates have the potential to address significant unmet medical needs and intend to develop them as efficiently and rapidly as possible. In 2018, we expect to report preliminary Phase 2 data for rilonacept and, if the Phase 2 data are favorable, we plan to initiate a Phase 3 clinical trial for rilonacept in recurrent pericarditis. For KPL-716, we anticipate reporting preliminary data from the single dose cohorts in our Phase 1a/1b clinical trial in normal healthy volunteers and subjects with atopic dermatitis in the second half of 2018. We also expect to initiate a Phase 2 clinical trial of mavrilimumab in GCA in 2018.

- **Commercialize our product candidates to bring new or improved therapies to patients in need.** We intend to market and commercialize our product candidates, if approved, in the United States and select international markets by developing our own sales, marketing, medical affairs and reimbursement organizations. We anticipate creating a targeted sales organization that supports specialist physicians who treat these specific patient populations and plan to build out this organization as our product candidates approach potential regulatory approval. We believe this approach will allow us to effectively reach patients and prescribers that our product candidates target and leverage the commercial potential of our product candidates.

- **Maximize our existing portfolio opportunity by expanding use across multiple indications.** A core component of our approach to product development is identifying assets that each have the potential to treat multiple diseases. We aim to develop and commercialize our product candidates to produce meaningful impact for patients across all relevant indications. Our assets are designed to specifically modulate signaling pathways that are implicated across a spectrum of autoimmune and autoinflammatory conditions. For example, our lead product candidate, rilonacept, is being studied in recurrent pericarditis, and we believe it may be effective in other IL-1a-mediated diseases characterized by painful serosal inflammation. We also believe that both mavrilimumab and KPL-716 have potential in additional indications.

- **Leverage our value-driven approach to identify, acquire, discover and develop new therapies.** We follow a disciplined and methodical approach to our review of new opportunities. We focus on research-based and comprehensive indication mapping exercises to categorize and prioritize indications of interest. We evaluate a variety of factors for potential product candidates and discovery targets, including biologic rationale for addressing the disease, potential for regulatory approval, commercial viability, intellectual property position, prospects for favorable pricing and reimbursement and the impact of competition. We also look at assets that could potentially address multiple indications. In building our current pipeline, we evaluated a large number of opportunities and negotiated agreements with parties for the assets that met our criteria and have acquired the rights to develop and commercialize five separate biologics. Going forward, we intend to be opportunistic in our business development activities.

- **Build our core capability in autoimmune and autoinflammatory diseases to establish a leadership position in the field.** Our current pipeline consists of protein therapeutics across various stages of drug development, including a cytokine trap, rilonacept, and four monoclonal antibodies—mavrilimumab, KPL-716, KPL-045 and KPL-404. Both categories of therapeutics functionally inhibit signaling pathways that are implicated in autoinflammatory-

or autoimmune-driven pathologies. We intend to leverage our internal discovery efforts and business development capabilities to complement our existing portfolio to build our core capability and establish a leadership position in the field.

Our Capital Structure

Following this offering, we will have four classes of common shares: Class A, Class A1, Class B and Class B1. All classes of our common shares will be economically equivalent to each other. The rights of the holders of our Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares will be identical, except with respect to voting, conversion and transferability. Holders of our Class A common shares — the only class of common shares being sold in this offering — will be entitled to one vote per Class A common share, while holders of our Class B common shares will be entitled to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares will have no associated voting rights. Following this offering, the Class A common shares will account for 22.2% of our aggregate voting power and the Class B common shares will account for the remaining 77.8% of the aggregate voting power. In addition, the number of Class A1 common shares to be outstanding after this offering will be 12,995,954 and the number of Class B1 common shares to be outstanding after this offering will be 16,057,618. See "Principal Shareholders" and "Description of Share Capital" for more information on beneficial ownership immediately following this offering.

As a result of the Class A common shares and Class B common shares that they will hold upon the closing of this offering, our executive officers and certain other members of our senior management will be able to exercise voting rights with respect to an aggregate of 856,166 Class A common shares and 4,244,005 Class B common shares, which will collectively represent 72.6% of the voting power of our outstanding share capital immediately following this offering. As a result, our executive officers and certain other members of our senior management will have the ability to control the outcome of all matters submitted to our shareholders for approval, including the election, removal, and replacement of directors and any merger, consolidation, or sale of all or substantially all of our assets. However, this percentage may change depending on any conversion of Class A1 common shares, Class B1 common shares or Class B common shares. Each holder of Class A1 common shares may elect to convert its Class A1 common shares into voting Class A common shares at any time, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of the issued and outstanding Class A common shares. Each holder of Class B1 common shares may elect to convert its Class B1 common shares into voting Class A common shares or Class B common shares at any time, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of the issued and outstanding Class A common shares. Any such holder of Class A1 common shares or Class B1 common shares will have the right to increase, decrease or waive this beneficial ownership limitation at its sole discretion by providing us with 61-days' prior written notice. As a result, upon 61-days' prior written notice, entities affiliated with one of our directors, Felix J. Baker, could convert their Class A1 common shares and Class B1 common shares into Class A common shares and Class B common shares, respectively, which in the aggregate would result in such entities holding 73.8% of the voting power of our outstanding share capital following this offering.

This concentrated control could delay, defer, or prevent a change of control, merger, consolidation, or sale of all or substantially all of our assets that our other shareholders support. Conversely, this concentrated control could allow our executive officers and certain other members of our senior management to consummate a transaction that our other shareholders do not support. See "Risk Factors — Risks Related to Our Common Shares and This Offering — After this

offering, our executive officers and certain other members of our senior management will have the ability to control all matters submitted to shareholders for approval."

Risks Associated with Our Business

Our business is subject to numerous risks and uncertainties, including those highlighted in the section titled "Risk Factors" immediately following this prospectus summary. Some of these risks are:

- we have a limited operating history, have never generated any product revenue, have incurred significant operating losses since our inception, expect to incur significant operating losses for the foreseeable future and may never achieve or maintain profitability;
- we may not be successful in our efforts to identify, discover, develop or acquire additional product candidates;
- we depend heavily on the success of our product candidates and cannot give any assurance that our product candidates will receive regulatory approval for any indication, which is necessary before they can be commercialized;
- we will need additional funding to complete the development and commercialization of our product candidates, if approved, and to acquire additional product candidates, and if we are unable to raise capital when needed, we could be forced to delay, reduce or eliminate our product development programs or future commercialization efforts;
- we had no involvement with or control over the pre-clinical and clinical development of our current product candidates prior to our acquisition of them, and we are dependent on the parties from whom we licensed or acquired such product candidates having conducted their research and development in accordance with the applicable protocols and standards, accurately reported the results of all clinical trials conducted prior to our acquisition and correctly collected and interpreted the data from these trials;
- we have acquired product candidates with positive clinical data in diseases other than our target indications, and we cannot be certain that our product candidates will prove to be effective in treating our target indications;
- we rely, and expect to continue to rely, on third parties to conduct our clinical trials and to manufacture our product candidates for pre-clinical and clinical testing, including our sole source of supply for each of our active pharmaceutical ingredients, and those third parties may not perform satisfactorily, which could delay our product development activities;
- all of our product candidates have been licensed or acquired from other parties; if we are unable to adequately protect our product candidates, or to secure and maintain freedom to operate, others could preclude us from commercializing our product candidates or compete against us more directly;
- we face significant competition from other biotechnology and pharmaceutical companies;
- concentration of ownership of the voting power of our common shares may prevent new investors in this offering from influencing significant corporate decisions; and
- we will likely be classified as a passive foreign investment company and we believe we have been classified as a controlled foreign corporation in the current taxable year and may be classified as a passive foreign investment company or controlled foreign corporation in any future taxable year, which may result in adverse U.S. federal income tax consequences to U.S. holders of our Class A common shares.

Our Corporate Information

We are an exempted company incorporated under the laws of Bermuda in July 2015. Our registered office is located in Bermuda at Clarendon House, 2 Church Street, Hamilton HM11, Bermuda. The telephone number of our registered office is +1 (441) 295-5950. Our website address is *www.kiniksa.com*. The information contained on our website is not incorporated by reference into this prospectus, and you should not consider any information contained on, or that can be accessed through, our website as part of this prospectus or in deciding whether to purchase our Class A common shares.

Implications of Being an Emerging Growth Company

We qualify as an "emerging growth company" as defined in Section 2(a) of the Securities Act of 1933, as amended, or the Securities Act, as modified by the Jumpstart Our Business Startups Act of 2012, or the JOBS Act. An "emerging growth company" may take advantage of exemptions from some of the reporting requirements that are otherwise applicable to public companies. These exceptions include:

- being permitted to present only two years of audited financial statements, in addition to any required unaudited interim financial statements, with correspondingly reduced "Management's Discussion and Analysis of Financial Condition and Results of Operations" disclosure in this prospectus;
- not being required to comply with the auditor attestation requirements in the assessment of our internal control over financial reporting;
- an exemption from compliance with the requirement of the Public Company Accounting Oversight Board regarding the communication of critical audit matters in the auditor's report on the financial statements;
- reduced disclosure obligations regarding executive compensation in our periodic reports, proxy statements and registration statements; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachute payments not previously approved.

We may take advantage of these exemptions until the last day of our fiscal year following the fifth anniversary of the closing of this offering. However, we will cease to be an emerging growth company prior to the end of such five-year period if (i) we become a "large accelerated filer" as defined in Rule 12b-2 under the Securities Exchange Act of 1934, as amended, which would occur if the market value of our common equity held by non-affiliates exceeds $700 million as of the last business day of our most recently completed second fiscal quarter; (ii) our annual gross revenue exceeds $1.07 billion; or (iii) we issue more than $1.0 billion of non-convertible debt in any three-year period.

We have elected to take advantage of certain of the reduced disclosure obligations in this prospectus and may elect to take advantage of other reduced reporting requirements in future filings. As a result, the information that we provide to our shareholders may be different than you might receive from other public reporting companies in which you hold equity interests.

In addition, the JOBS Act provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. We have irrevocably elected not to avail ourselves of this exemption and, therefore, we will be subject to the same new or revised accounting standards as other public companies that are not emerging growth companies.

The Offering

Class A common shares offered by us	7,000,000 shares
Option to purchase additional Class A common shares	1,050,000 shares
Class A common shares to be outstanding after this offering	13,270,569 shares (14,320,569 shares if the underwriters exercise their option to purchase additional Class A common shares in full)
Class B common shares to be outstanding after this offering	4,638,855 shares
Class A1 common shares to be outstanding after this offering	12,995,954 shares
Class B1 common shares to be outstanding after this offering	16,057,618 shares
Total common shares to be outstanding after this offering	46,962,996 shares (48,012,996 shares if the underwriters exercise their option to purchase additional Class A common shares in full)
Voting rights	Following this offering, we will have four classes of common shares outstanding: Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares. Each Class A common share will entitle its holder to one vote per Class A common share. Each Class B common share will entitle its holder to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares will not have voting rights. Immediately following this offering, the holders of our Class A common shares will account for 22.2% of our aggregate voting power and the holders of our Class B common shares will account for the remaining 77.8% of our aggregate voting power. See "Principal Shareholders" and "Description of Share Capital" for additional information.
Use of proceeds	We estimate that the net proceeds from this offering will be approximately $113.4 million (or approximately $131.0 million if the underwriters exercise their option to purchase additional Class A common shares in full), based on an assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus.

	We intend to use the net proceeds from this offering to advance the clinical development of rilonacept, mavrilimumab and KPL-716, and to fund other research and development activities and for working capital and general corporate purposes. See "Use of Proceeds" beginning on page 83.
Risk factors	See "Risk Factors" beginning on page 13 and the other information included in this prospectus for a discussion of factors you should consider carefully before deciding to invest in our Class A common shares.
Proposed Nasdaq Global Market symbol	"KNSA"

The total number of common shares to be outstanding after this offering is based on 6,270,569 Class A common shares and 4,638,855 Class B common shares outstanding as of March 31, 2018 and includes the conversion of all of our preferred shares outstanding as of March 31, 2018 into 5,546,019 Class A common shares, 1,070,502 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares, in each case upon the closing of this offering. This amount excludes:

- 4,702,190 Class A common shares issuable upon exercise of share options outstanding as of March 31, 2018, at a weighted average exercise price of $5.41 per share;
- 4,466,500 Class A common shares that will become available for future issuance under our 2018 Incentive Award Plan, or 2018 Plan, which will become effective in connection with this offering, as well as common shares that become available pursuant to provisions in the 2018 Plan that automatically increase the share reserve under the 2018 Plan as described in "Executive and Director Compensation — Executive Compensation Plans — 2018 Incentive Award Plan" (including 122,971 Class A common shares issuable upon exercise of share options that will be granted to certain of our directors and employees, at an exercise price per share equal to the initial public offering price of our Class A common shares in this offering); and
- 670,000 Class A common shares that will become available for future issuance under our 2018 Employee Share Purchase Plan, or the ESPP, which will become effective in connection with this offering, as well as common shares that become available pursuant to provisions in the ESPP that automatically increase the share reserve under the ESPP as described in "Executive and Director Compensation — Executive Compensation Plans — 2018 Employee Share Purchase Plan".

Unless otherwise indicated, this prospectus reflects and assumes the following:

- a 1-for-2.73235 reverse share split of our common and preferred shares effected on May 11, 2018;
- the conversion of all of our preferred shares into 5,546,019 Class A common shares, 12,995,954 Class A1 common shares, 1,070,502 Class B common shares and 16,057,618 Class B1 common shares, in each case upon the closing of this offering;
- no exercise of outstanding share options after March 31, 2018;
- the effectiveness of our amended and restated bye-laws immediately following the closing of this offering; and

- no exercise by the underwriters of their option to purchase additional Class A common shares.

Certain of our existing shareholders, including shareholders affiliated with one of our directors, have indicated an interest in purchasing an aggregate of approximately $50.0 million of Class A common shares in this offering at the initial public offering price per share. However, because indications of interest are not binding agreements or commitments to purchase, the underwriters may determine to sell more, less or no shares in this offering to any of these existing shareholders, or any of these existing shareholders may determine to purchase more, less or no shares in this offering. The underwriters will receive the same underwriting discount on any shares purchased by these existing shareholders as they will on any other shares sold to the public in this offering.

SUMMARY CONSOLIDATED FINANCIAL DATA

You should read the following summary consolidated financial data together with our consolidated financial statements and the related notes appearing at the end of this prospectus and the "Selected Consolidated Financial Data" and "Management's Discussion and Analysis of Financial Condition and Results of Operations" sections of this prospectus. We have derived the consolidated statement of operations data for the years ended December 31, 2016 and 2017 from our audited consolidated financial statements appearing at the end of this prospectus. We have derived the consolidated statement of operations data for the three months ended March 31, 2017 and 2018 and the consolidated balance sheet data as of March 31, 2018 from our unaudited consolidated financial statements appearing at the end of this prospectus, which have been prepared on the same basis as the audited consolidated financial statements. In the opinion of management, the unaudited data reflects all adjustments, consisting only of normal recurring adjustments, necessary for a fair statement of the financial information in those statements. Our historical results are not necessarily indicative of results that may be expected in any future period, and our results for any interim period are not necessarily indicative of results that may be expected for any full year.

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
	(in thousands, except share and per share data)			
Consolidated Statement of Operations Data:				
Operating expenses:				
Research and development	$ 17,439	$ 56,357	$ 3,145	$ 12,630
General and administrative	6,563	9,043	1,903	3,710
Total operating expenses	24,002	65,400	5,048	16,340
Loss from operations	(24,002)	(65,400)	(5,048)	(16,340)
Interest income	65	529	74	305
Loss before benefit (provision) for income taxes	(23,937)	(64,871)	(4,974)	(16,035)
Benefit (provision) for income taxes	(36)	(2)	33	53
Net loss	$ (23,973)	$ (64,873)	$ (4,941)	$ (15,982)
Net loss per share attributable to common shareholders—basic and diluted[1]	$ (91.61)	$ (35.85)	$ (3.52)	$ (6.45)
Weighted average common shares outstanding—basic and diluted[1]	261,695	1,809,751	1,405,400	2,478,903
Pro forma net loss per share attributable to common shareholders—basic and diluted (unaudited)[1]		$ (2.74)		$ (0.49)
Pro forma weighted average common shares outstanding—basic and diluted (unaudited)[1]		23,638,410		32,466,951

(1) See Note 11 to our consolidated financial statements included elsewhere in this prospectus for further details on the calculation of basic and diluted net loss per share attributable to common shareholders and on the calculation of pro forma basic and diluted net loss per share attributable to common shareholders.

	As of March 31, 2018		
	Actual	Pro Forma[2]	Pro Forma As Adjusted[3]
	(in thousands)		
Consolidated Balance Sheet Data:			
Cash and cash equivalents	$ 221,108	$ 221,108	$ 334,517
Working capital[1]	203,185	203,185	318,173
Total assets	224,775	224,775	336,576
Convertible preferred shares	310,592	—	—
Total shareholders' (deficit) equity	(105,115)	205,477	318,857

(1) We define working capital as current assets less current liabilities.

(2) The pro forma balance sheet data give effect to the conversion of all of our outstanding preferred shares into an aggregate of 35,670,093 common shares upon the closing of this offering.

(3) The pro forma as adjusted balance sheet data give further effect to our issuance and sale of 7,000,000 Class A common shares in this offering at an assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

Each $1.00 increase (decrease) in the assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, working capital, total assets and total shareholders' equity by $6.5 million, assuming that the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us. Each increase (decrease) of 1,000,000 shares in the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, working capital, total assets and total shareholders' equity by $16.7 million, assuming no change in the assumed initial public offering price per share and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

RISK FACTORS

Investing in our Class A common shares involves a high degree of risk. You should carefully consider the risks described below, as well as the other information in this prospectus, including our consolidated financial statements and the related notes and "Management's Discussion and Analysis of Financial Condition and Results of Operations," before deciding whether to invest in our Class A common shares. The occurrence of any of the events or developments described below could adversely affect our business, financial condition, results of operations and growth prospects. In such an event, the market price of our Class A common shares could decline, and you may lose all or part of your investment. Additional risks and uncertainties not presently known to us or that we currently deem immaterial also may impair our business operations.

Risks Related to Our Financial Position and Capital Needs

We are a clinical-stage biopharmaceutical company with a limited operating history and have not generated any revenue from product sales. We have incurred significant operating losses since our inception and anticipate that we will incur continued losses for the foreseeable future.

We have incurred losses in each year since our inception in 2015 and anticipate incurring losses for the foreseeable future. To date, we have invested substantially all of our efforts and financial resources in identifying, acquiring, in-licensing and developing our product candidates, including commencing and conducting clinical trials and providing general and administrative support for these operations. Our future success is dependent on our ability to develop, obtain regulatory approval for and successfully commercialize one or more of our product candidates. We have not yet demonstrated our ability to initiate or successfully complete any Phase 3 or other pivotal clinical trials, obtain regulatory approvals, manufacture a commercial scale drug, or conduct sales and marketing activities. We currently generate no revenue from sales of any products, and we may never be able to develop or commercialize a marketable product. Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk. Typically, it takes many years to develop one new drug from the time it is discovered to when it is available for treating patients, and development may cease for a number of reasons. Consequently, predictions about our future success or viability could be more accurate if we had a longer operating history.

We have incurred significant losses related to expenses for research and development and our ongoing operations. Our net losses for the years ended December 31, 2016 and 2017 and three months ended March 31, 2017 and 2018 were $24.0 million, $64.9 million, $4.9 million and $16.0 million, respectively. As of March 31, 2018, we had an accumulated deficit of $107.0 million. We expect to continue to incur losses for the foreseeable future, and we anticipate these losses will increase substantially as we:

- continue our research and pre-clinical and clinical development of our product candidates, including our ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept for the treatment of recurrent pericarditis and our ongoing Phase 1a/1b clinical trial of KPL-716 in healthy volunteers and in subjects with atopic dermatitis, and commence our Phase 2 clinical trial of mavrilimumab for the treatment of GCA;
- expand the scope of our current clinical trials for our product candidates;
- advance our programs into more expensive clinical trials, including our plans to commence a Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis;
- initiate additional pre-clinical studies and clinical trials for our product candidates;

- increase our manufacturing needs or add additional manufacturers or suppliers;
- seek regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;
- establish a sales, marketing and distribution infrastructure to commercialize any products for which we may obtain marketing approval;
- seek to identify, assess, acquire or develop additional product candidates;
- make milestone or other payments under any license or purchase agreements;
- seek to maintain, protect and expand our intellectual property portfolio;
- seek to attract and retain skilled personnel;
- create additional infrastructure to support our operations as a public company and our product development and planned future commercialization efforts; and
- experience any delays or encounter issues with any of the above, including but not limited to failed trials, complex results, safety issues, other regulatory challenges that require longer follow-up of existing trials, additional major trials or additional supportive trials in order to pursue marketing approval.

Further, the net losses we incur may fluctuate significantly from quarter-to-quarter and year to year, such that a period to period comparison of our results of operations may not be a good indication of our future performance. Once we are a public company, we will incur additional costs associated with operating as a public company. Even if we achieve profitability in the future, we may not be able to sustain profitability in subsequent periods. Our prior losses, combined with expected future losses, have had and will continue to have an adverse effect on our shareholders' equity and working capital.

We will require substantial additional financing, and a failure to obtain this necessary capital when needed on acceptable terms, or at all, could force us to delay, limit, reduce or terminate our product development, other operations or commercialization efforts.

The development and commercialization of biopharmaceutical products is capital intensive. We are advancing our product candidates through pre-clinical and clinical development and, in 2018, anticipate beginning new clinical trials for our product candidates, rilonacept, mavrilimumab and KPL-716. We expect our expenses to increase in connection with our ongoing activities as we continue the research and development of, and, if successful, seek marketing approval for, our product candidates. In addition, if we obtain marketing approval for any of our product candidates, we expect to incur significant commercialization expenses related to manufacturing, product sales, marketing, and distribution. As our product candidates progress through development and towards commercialization, we will need to make milestone payments to the licensors and other third parties from whom we have acquired our product candidates. We may also need to raise additional funds sooner if we choose to pursue additional indications for our product candidates or otherwise expand more rapidly than we presently anticipate. Furthermore, upon the closing of this offering, we expect to incur additional costs associated with operating as a public company. Accordingly, we will need to obtain substantial additional funding in connection with our continuing operations. If we are unable to raise capital when needed on attractive terms, if at all, we will be forced to delay, reduce or eliminate certain of our clinical development plans, research and development programs or future commercialization efforts.

The development process for our product candidates is highly uncertain, and we cannot estimate with certainty the actual amounts necessary to successfully complete the development,

regulatory approval process and commercialization of our product candidates. Our operating plans may change as a result of many factors currently unknown to us, and we may need to seek additional funds sooner than expected, through public or private equity, debt financings or other sources. Our future capital requirements will depend on and could increase significantly as a result of many factors, including:

- the results, time and cost necessary for completing our open-label Phase 2 proof-of-concept clinical trial of rilonacept for the treatment of recurrent pericarditis and our Phase 1a/1b clinical trials of KPL-716 for the treatment of atopic dermatitis and commencing our planned Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, our planned Phase 2 clinical trial for mavrilimumab for the treatment of GCA and our planned Phase 2 clinical trial for KPL-716 for the treatment of prurigo nodularis;
- the number, size and type of any additional clinical trials;
- the costs, timing and outcomes of seeking and potentially obtaining approvals from the FDA or comparable foreign regulatory authorities, including the potential for the FDA or comparable regulatory authorities to require that we conduct more studies than those that we currently expect to conduct and the costs of post-marketing studies or risk evaluation and mitigation strategies, or REMS, that could be required by regulatory authorities;
- the costs and timing of transferring manufacturing technology to third-party manufacturers, producing product candidates to support clinical trials and preparing to manufacture mavrilimumab and KPL-716 on a commercial scale, as well as producing rilonacept in potential new final form configurations;
- the timing and amount of milestone or other payments we must make under our agreements with Regeneron Pharmaceuticals, Inc., or Regeneron, MedImmune, Limited, or MedImmune, Biogen MA Inc., or Biogen, Novo Nordisk A/S, or Novo Nordisk, and the other third parties from whom we have acquired or in-licensed our product candidates or from whom we may in the future acquire or in-license product candidates or in connection with the exercise of our option to purchase all of the outstanding capital stock of Primatope Therapeutics, Inc., or Primatope;
- our ability to successfully commercialize any of our product candidates, including the cost and timing of forming and expanding our sales organization and marketing capabilities;
- the amount of sales revenues from our product candidates, if approved, including the sales price and the availability of coverage and adequate third-party reimbursement;
- competitive and potentially competitive products and technologies and patients' receptivity to our product candidates and the technology underlying them in light of competitive products and technologies;
- the cash requirements of any future acquisitions, developments or discovery of additional product candidates, including any licensing or collaboration agreements;
- the time and cost necessary to respond to technological and market developments;
- the costs of filing, prosecuting, defending and enforcing any patent claims and other intellectual property rights;
- any product liability or other lawsuits related to our product candidates or any products;
- the costs associated with being a public company;
- our need and ability to hire additional personnel; and

- the receptivity of the capital markets to financings by biotechnology companies generally and companies with product candidates and technologies such as ours specifically.

Any additional fundraising efforts may divert our management from their day-to-day activities, which may adversely affect our ability to develop and commercialize our product candidates. Dislocations in the financial markets may make equity and debt financing more difficult to obtain, and may have a material adverse effect on our ability to meet our fundraising needs when they arise. Additional funds may not be available when we need them, on terms that are acceptable to us, or at all. If we are unable to obtain funding on a timely basis, we may be required to significantly curtail, delay or discontinue one or more of our pre-clinical studies, clinical trials or other research or development programs, the commercialization of any product candidate. We may also be unable to expand our operations or otherwise capitalize on our business opportunities or may be required to relinquish rights to our product candidates or products. Any of these occurrences could materially affect our business, financial condition and results of operations.

Raising additional capital may cause dilution to our shareholders, including purchasers of shares in this offering, restrict our operations or require us to relinquish rights to our technologies or product candidates.

Until such time as we can generate substantial product revenues, if ever, we expect to finance our cash needs through securities offerings or debt financings, or possibly, license and collaboration agreements or research grants. The terms of any financing may adversely affect the holdings or the rights of our shareholders and our issuance of additional securities, whether equity or debt, or the possibility of such issuance, may cause the market price of our Class A common shares to decline. The sale of additional equity or convertible securities would dilute all of our shareholders, including your ownership interest. The incurrence of indebtedness would result in increased fixed payment obligations and we may be required to agree to certain restrictive covenants, such as limitations on our ability to incur additional debt, limitations on our ability to acquire, sell or license intellectual property rights and other operating restrictions that could adversely impact our ability to conduct our business. We could also be required to seek funds through arrangements with collaborators or otherwise at an earlier stage than otherwise would be desirable and we may be required to relinquish rights to some of our technologies, product candidates or future revenue streams, or otherwise agree to terms unfavorable to us, any of which may have a material adverse effect on our business, operating results and prospects. If we raise funds through research grants, we may be subject to certain requirements, which may limit our ability to use the funds or require us to share information from our research and development. Raising additional capital through any of these or other means could adversely affect our business and the holdings or rights of our shareholders, and may cause the market price of our shares to decline.

Risks Related to Product Development and Regulatory Approval

We depend heavily on the success of rilonacept, mavrilimumab and KPL-716, which are in various stages of clinical development. If we are unable to advance our product candidates in clinical development, obtain regulatory approval and ultimately commercialize our product candidates, or experience significant delays in doing so, our business will be materially harmed.

We do not currently generate any revenue from sales of any products, and we may never be able to develop or commercialize marketable products. Each of our product candidates require additional clinical development, management of pre-clinical, clinical or manufacturing activities, regulatory approval, obtaining adequate manufacturing supply, building of a commercial organization, and significant marketing efforts before we generate any revenue from product sales.

We have three product candidates in various stages of clinical development and two at the pre-clinical development stage. None of them have been previously studied in the indications for which we are developing them. We may not be able to demonstrate that they are safe or effective in the indications for which we are studying them and they may not be approved. Although rilonacept is approved and marketed for human use for the treatment of CAPS in the United States by Regeneron, we are studying rilonacept for the treatment of recurrent pericarditis in an open-label Phase 2 proof-of-concept clinical trial, and, if the preliminary data are favorable, we plan to advance development to a Phase 3 clinical trial in 2018. Mavrilimumab has been through Phase 2 clinical trials conducted by MedImmune for the treatment of rheumatoid arthritis, or RA, but we plan to enter into Phase 2 clinical trials with mavrilimumab for the treatment of GCA. Our third product candidate, KPL-716, is currently undergoing a Phase 1a clinical trial in healthy volunteers and a Phase 1b clinical trial in subjects with atopic dermatitis and, if the data from our Phase 1a/1b clinical trial are favorable, we intend to commence Phase 2 clinical trials for atopic dermatitis as well as prurigo nodularis. Our assumptions about why these product candidates are worthy of future development and potential approval in these, or any, indications are based on indirect data primarily collected by other companies. We also have pre-clinical product candidates that will need to progress through IND-enabling studies prior to clinical development. None of our product candidates have advanced into a pivotal study for the indications for which we are studying. We are not permitted to market or promote any of our product candidates before we receive regulatory approval from the FDA or comparable foreign regulatory authorities.

We have not submitted, and we may never submit marketing applications to the FDA or comparable foreign regulatory authorities for our product candidates. We cannot be certain that any of our product candidates will be successful in clinical trials or receive regulatory approval. Further, our product candidates may not receive regulatory approval even if they are successful in clinical trials. If we do not receive regulatory approvals for one or more of our product candidates, we may not be able to continue our operations.

Each of our product candidates will require additional pre-clinical and/or clinical development, regulatory approval in one or more jurisdictions, obtaining manufacturing supply, capacity and expertise, building of a commercial organization, substantial investment and significant marketing efforts before we are able to generate any revenue from product sales. The success of our product candidates will depend on several factors, including the following:

- successful completion of pre-clinical studies, including toxicology studies, biodistribution studies and minimally efficacious dose studies in animals, conducted, where applicable, under the FDA's Good Laboratory Practice, or GLP, regulations;
- submission to the FDA of INDs and of clinical trial applications to foreign governmental authorities, for our product candidates to commence planned clinical trials or future clinical trials;
- successful enrollment in, and completion of, clinical trials, the design and implementation of which are agreed to by the applicable regulatory authorities, and the conduct of clinical trials by contract research organizations, or CROs, to successfully conduct such trials within our planned budget and timing parameters and without materially adversely impacting our trials;
- successful data from our clinical programs that support an acceptable risk-benefit profile of our product candidates for the targeted indications in the intended populations to the satisfaction of the applicable regulatory authorities;
- timely receipt, if at all, of regulatory approvals from applicable regulatory authorities;

- establishment of arrangements with third-party manufacturers, as applicable, for continued clinical supply and commercial manufacturing;
- successful development of our manufacturing processes and transfer to new third-party facilities to support future development activities and commercialization that are operated by contract manufacturing organizations, or CMOs, in a manner compliant with all regulatory requirements;
- establishment and maintenance of patent and trade secret protection or regulatory exclusivity for our product candidates;
- successful commercial launch of our product candidates, if and when approved;
- acceptance of our products, if and when approved, by patients, the medical community and third-party payors;
- effective competition with other therapies;
- establishment and maintenance of adequate healthcare coverage and reimbursement;
- enforcement and defense of intellectual property rights and claims;
- continued compliance with any post-marketing requirements imposed by regulatory authorities, including any required post-marketing clinical trials or REMS; and
- maintenance of a continued acceptable safety profile of the product candidates following approval.

If we do not accomplish one or more of these factors in a timely manner or at all, we could experience significant delays or an inability to successfully commercialize our product candidates, which would materially harm our business. If we do not receive regulatory approvals for one or more of our product candidates, we may not be able to continue our operations. Even if we successfully obtain regulatory approvals to manufacture and market our product candidates, our revenues will be dependent, in part, upon the size of the markets in the territories for which we gain regulatory approval and have commercial rights. If the markets for patient subsets that we are targeting are not as significant as we estimate, we may not generate significant revenues from sales of such products, if approved.

We plan to seek regulatory approval to commercialize our product candidates in the United States and potentially in foreign countries. While the scope of regulatory approval is similar in many countries, to obtain separate regulatory approval in multiple countries will require us to comply with numerous and varying regulatory requirements of each such country or jurisdiction regarding safety and efficacy and governing, among other things, clinical trials and commercial sales, pricing and distribution, and we cannot predict success in any such jurisdictions.

Clinical drug development is a lengthy and expensive process with uncertain timelines and uncertain outcomes. We may encounter substantial delays in our clinical trials, or we may fail to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities. We may therefore be unable to obtain required regulatory approvals and be unable to commercialize our product candidates on a timely basis, if at all.

Before obtaining marketing approval from regulatory authorities for the sale of our product candidates, we must conduct extensive clinical trials to demonstrate the safety and efficacy of the product candidates in humans. Clinical testing is expensive, time consuming and uncertain as to outcome. We cannot guarantee that any clinical trials will be conducted as planned or completed on schedule, if at all.

Subject to obtaining favorable preliminary data from our ongoing open-label Phase 2 proof-of-concept clinical trial of rilonacept, we plan to initiate a Phase 3 clinical trial for rilonacept as a treatment for recurrent pericarditis in 2018. We have not yet had any discussions with the FDA regarding the design of a Phase 3 clinical trial for rilonacept for treatment of recurrent pericarditis. We also plan to initiate a Phase 2 clinical trial of mavrilimumab for the treatment of GCA in 2018. Subject to favorable data from our Phase 1a/1b clinical trial of KPL-716 in healthy volunteers and subjects with atopic dermatitis, we plan to commence Phase 2 clinical trials of KPL-716 for the treatment of atopic dermatitis as well as prurigo nodularis. We are also continuing preparation for IND-enabling studies of KPL-045 and KPL-404 prior to initiating clinical trials. Commencing our planned clinical trials is subject to acceptance by the FDA of an IND or an IND amendment, or acceptance by European regulatory authorities of a CTA, as applicable, and finalizing the trial design based on discussions with the FDA, European regulatory authorities or other applicable regulatory authorities. Even after we receive and incorporate guidance from these regulatory authorities, the FDA or other regulatory authorities could disagree that we have satisfied their requirements to commence our clinical trials, disagree with our interpretation of data from the relevant pre-clinical studies, clinical trials or CMC data, or disagree or change their position on the acceptability of our trial designs including the proposed dosing schedule, our definitions of the patient populations or the clinical endpoints selected, which may require us to complete additional pre-clinical studies, clinical trials, CMC development, other studies or impose stricter approval conditions than we currently expect. For example, prior to us licensing mavrilimumab, MedImmune submitted an IND to the FDA to conduct a clinical trial of mavrilimumab in RA, and the FDA issued a clinical hold based on its review of certain effects in the lungs observed in non-human primates in pre-clinical toxicity studies. However, following subsequent discussions between MedImmune and the FDA regarding the clinical hold and the availability of additional clinical safety data that MedImmune generated in human clinical trials conducted outside of the United States subsequent to the original IND submission, the FDA acknowledged that the risk/benefit assessment for investigation of mavrilimumab in a clinical trial may differ depending on the patient population studied. Specifically, the FDA acknowledged that the risk/benefit assessment for initiation of a clinical trial may be considered favorable in a patient population with high morbidity and limited effective treatment options, including refractory RA. We believe that the FDA's communications with MedImmune suggest that the FDA could find an acceptable risk/benefit for a clinical trial of mavrilimumab in the United States in GCA, a disease with high morbidity and limited treatment options, which we are pursuing. However, the FDA may disagree and may require that we generate additional data, or that we implement additional monitoring or other trial design changes prior to initiating clinical trials of mavrilimumab in the United States.

Further, we could discover that our clinical trial design leads to enrollment difficulties which could require protocol amendments and further delay our study. Successful completion of our clinical trials is a prerequisite to submitting a biologics license application, or BLA, to the FDA and a marketing authorization application, or MAA, to the European Medicines Agency, or EMA, for each product candidate and, consequently, to obtaining approval and initiating commercial marketing of our current and future product candidates. We do not know whether any of our clinical trials will begin or be completed on schedule, if at all.

A failure of one or more clinical trials can occur at any stage of testing, and our future clinical trials may not be successful. We may experience delays in our ongoing clinical trials and we do not know whether planned clinical trials will begin on time, will be allowed by regulatory authorities, need to be redesigned, enroll patients on time or will be completed on schedule, if at all. Events

that may prevent successful or timely completion of clinical development include but are not limited to:

- inability to generate sufficient pre-clinical, toxicology or other in vivo or in vitro data to support the initiation of human clinical trials;
- delays in reaching a consensus with regulatory agencies on trial design or implementation;
- delays in establishing the appropriate dosage levels or frequency of dosing or treatment in clinical trials;
- delays in reaching, or failing to reach, agreement on acceptable terms with prospective CROs, and clinical trial sites, the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and clinical trial sites;
- difficulties in obtaining required Institutional Review Board, or IRB, approval at each clinical trial site;
- delays in or failure to obtain regulatory approval to commence a trial, or imposition of a clinical hold by regulatory agencies, after review of an IND, application or amendment, or equivalent application or amendment, or an inspection of our clinical trial operations or study sites;
- challenges in recruiting and enrolling suitable patients to participate in our clinical trials;
- amendments to protocols amending study criteria and design;
- difficulty collaborating with patient groups and investigators;
- failure by our CROs, other third parties, or us to adhere to clinical trial requirements or to perform their obligations in a timely or compliant manner;
- failure to perform in accordance with the FDA's good clinical practices requirements, or GCPs, or applicable regulatory guidelines in other countries;
- patients not completing participation in a clinical trial or return for post-treatment follow-up;
- clinical trial sites or patients dropping out of a clinical trial;
- participating patients experiencing serious adverse events or undesirable side effects or are exposed to unacceptable health risks;
- safety issues, including occurrence of adverse events associated with the product candidate that are viewed to outweigh its potential benefits;
- difficulty in identifying the populations that we are trying to enroll in a particular trial, which may delay enrollment and reduce the power of a clinical trial to detect statistically significant results;
- changes in regulatory requirements, policies and guidance that require amending or submitting new clinical protocols;
- the cost of clinical trials of our product candidates being greater than we anticipate;
- clinical trials of our product candidates producing negative or inconclusive results, which may result in us deciding, or regulators requiring us, to conduct additional clinical trials or abandon drug development programs; and
- delays in manufacturing, testing, releasing, validating or importing/exporting sufficient stable quantities of our product candidates for use in clinical trials or the inability to do any of the foregoing.

We could encounter delays if a clinical trial is suspended or terminated by us, the IRBs of the institutions in which such trials are being conducted, the Data Safety Monitoring Board, for such trial or the FDA or other regulatory authorities. Such authorities may impose such a suspension or termination due to a number of factors, including failure to conduct the clinical trial in accordance with regulatory requirements or our clinical protocols, inspection of the clinical trial operations or trial site by the FDA or other regulatory authorities resulting in the imposition of a clinical hold, unforeseen safety issues or adverse side effects that arise in our trial, failure to demonstrate a benefit from using a drug, changes in governmental regulations or administrative actions or lack of adequate funding to continue the clinical trial. Many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates.

Moreover, principal investigators for our clinical trials may serve as scientific advisors or consultants to us from time to time and receive compensation in connection with such services. Under certain circumstances, we may be required to report some of these relationships to the FDA or other regulatory authority. The FDA or other regulatory authority may conclude that a financial relationship between us and a principal investigator has created a conflict of interest or otherwise affected interpretation of the study. The FDA or other regulatory authority may therefore question the integrity of the data generated at the applicable clinical trial site and the utility of the clinical trial itself may be jeopardized. This could result in a delay in approval, or rejection, of our marketing applications by the FDA or other regulatory authority, as the case may be, and may ultimately lead to the denial of marketing approval of our product candidates.

If we experience delays in the completion of any clinical trial of our product candidates or any clinical trial of our product candidates is terminated, the commercial prospects of our product candidates may be harmed, and our ability to generate product revenues from our product candidates, if any, will be delayed. Moreover, any delays in completing our clinical trials will increase our costs, slow down the development and approval process of our product candidates and jeopardize our ability to commence product sales and generate revenue, if any. Significant clinical trial delays could also allow our competitors to bring products to market before we do or shorten any periods during which we have the exclusive right to commercialize our product candidates and could impair our ability to commercialize our product candidates. In addition, many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates.

Clinical trials must be conducted in accordance with the laws and regulations of the FDA, European Union rules and regulations and other applicable regulatory authorities' legal requirements, regulations or guidelines, and are subject to oversight by these governmental agencies and IRBs at the medical institutions where the clinical trials are conducted. In addition, clinical trials must be conducted with supplies of our product candidates produced under current good manufacturing practice, or cGMP, requirements and other regulations. Furthermore, we rely on CROs and clinical trial sites to ensure the proper and timely conduct of our clinical trials and while we have agreements governing their committed activities, we have limited influence over their actual performance. We depend on our collaborators and on medical institutions and CROs to conduct our clinical trials in compliance with GCP requirements. To the extent our collaborators or the CROs fail to enroll participants for our clinical trials, fail to conduct the study to GCP standards or are delayed for a significant time in the execution of trials, including achieving full enrollment, we may be affected by increased costs, program delays or both. In addition, clinical trials that are conducted in countries outside the European Union and the United States may subject us to further delays and expenses as a result of increased shipment costs, additional regulatory requirements and the engagement of non-EU and non-U.S. CROs, as well as expose us to risks associated with

clinical investigators who are unknown to the FDA or the EMA, and different standards of diagnosis, screening and medical care.

Further, conducting clinical trials in foreign countries, as we may do for certain of our product candidates, presents additional risks that may delay completion of our clinical trials. These risks include the failure of enrolled patients in foreign countries to adhere to clinical protocol as a result of differences in healthcare services or cultural customs, managing additional administrative burdens associated with foreign regulatory schemes, as well as political and economic risks relevant to such foreign countries.

We must produce, through third parties, sufficient stable quantities of our product candidates for use in our clinical trials. Any delays in the production of our product candidates may lead to a delay in our clinical trials. If we make manufacturing or formulation changes to our product candidates or change manufacturers or manufacturing processes, we may be unsuccessful in producing the product as compared to the process or manufacturer used in prior clinical trials, and therefore may need to conduct additional trials to bridge our modified product candidates to earlier versions, which could impact the timing of commencing or completing our clinical trials. Moreover, there is no assurance that future clinical trials utilizing a new formulation of a product candidate manufactured by different manufacturers or pursuant to a new process will result in the favorable result observed in the prior clinical trials of such product candidates as we have observed to date. For example, we will need to produce mavrilimumab using different media and feed compared to the processes that were used by MedImmune to develop our existing inventory. Further, we will need to identify a third party to manufacture mavrilimumab for any Phase 3 clinical trials and commercialization efforts, if any, and will need to transfer the manufacturing process of mavrilimumab to such third-party CMOs. This manufacturer may be unsuccessful in producing the product in quantities or quality necessary to support our clinical trials or commercialization efforts, if any, which would delay development of the mavrilimumab.

Clinical trial delays could also shorten any periods during which our products have patent protection and may allow our competitors to bring products to market before we do, which could impair our ability to obtain orphan exclusivity and to successfully commercialize our product candidates and may harm our business and results of operations. Any inability to successfully complete pre-clinical and clinical development could result in additional costs to us or impair our ability to generate revenue and harm our business, financial condition and prospects significantly.

We may find it difficult to enroll patients in our clinical trials in a timely manner given the limited number of patients who have the diseases for which our product candidates are being studied, as well as particular enrollment criteria. Difficulty in enrolling patients could delay or prevent clinical trials of our product candidates, and our research and development efforts could be adversely affected.

Identifying and qualifying patients to participate in clinical trials of our product candidates is critical to our success. The timing of our clinical trials depends in part on the speed at which we can recruit patients to participate in testing our product candidates, and we may experience delays in our clinical trials if we encounter difficulties in enrollment. Patient enrollment depends on many factors, including the size and nature of the patient population, eligibility criteria for the clinical trial, the proximity of patients to clinical sites, the design of the clinical protocol, the availability of competing clinical trials, the availability of new drugs approved for the indication the clinical trial is investigating, the risk that patients enrolled in clinical trials will drop out of the trials before completion of their treatment and clinicians' and patients' perceptions as to the safety and potential advantages of the product candidate being studied in relation to other available therapies.

Many of the conditions for which we plan to evaluate our current product candidates in the near future are in small disease populations. Accordingly, there are limited patient pools from which to draw for clinical trials.

In addition to the rarity of these diseases, the eligibility criteria of our clinical trials in any of our clinical trials will further limit the pool of available trial participants as we will require patients to have specific characteristics that we can measure or to assure their disease is either severe enough or not too advanced to include them in a trial. Further, we could learn that our clinical trial design increased the difficulty to enroll patients and could delay our trials. The process of finding and diagnosing patients may prove costly, especially since the rare diseases we are studying are commonly under diagnosed. We also may not be able to identify, recruit, enroll and retain a sufficient number of patients to complete our clinical trials because of the perceived risks and benefits of the product candidate under trial, the proximity and availability of clinical trial sites for prospective patients and the patient referral practices of physicians. The availability and efficacy of competing therapies and clinical trials, and clinicians' and patients' perceptions as to the potential advantages of the product candidate being studied in relation to those available competing therapies and clinical trials, can also adversely impact enrollment. If patients are unwilling to participate in our trials for any reason, the timeline for recruiting patients, conducting trials and obtaining regulatory approval of potential products may be delayed, the commercial prospects of our product candidates will be harmed, and our ability to generate product revenue from any of these product candidates could be delayed or prevented. Moreover, failure to obtain and maintain patient consents can also lead to delay or prevent completion of clinical trials of our product candidates.

In addition, our clinical trials may compete with other clinical trials for product candidates that are in the same therapeutic areas as our product candidates, and this competition may further reduce the number and types of patients available to us, because some patients who might have opted to enroll in our trials may instead opt to enroll in a trial being conducted by one of our competitors. Since the number of qualified clinical investigators is limited, we may conduct some of our clinical trials at the same clinical trial sites that some of our competitors use, which will reduce the number of patients who are available for our clinical trials in such clinical trial site. Delays in patient enrollment will increase our costs, slow down our product candidate development and approval process and jeopardize our ability to commence product sales and generate revenue. Any of these occurrences may harm our business, financial condition and prospects significantly. In addition, many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates.

Our product candidates may cause undesirable side effects or have other safety risks that could delay or prevent their regulatory approval, limit the commercial profile of an approved label or result in significant negative consequences, including withdrawal of approval, following any potential marketing approval.

Treatment with our product candidates may produce undesirable side effects or adverse reactions or events. Undesirable side effects caused by our product candidates could cause us or regulatory authorities to interrupt, delay or halt clinical trials and could result in a more restrictive label or the delay or denial of regulatory approval by the FDA or other comparable foreign authorities.

All of our product candidates modulate the immune system and carry risks associated with immunosuppression, including the theoretical risk of serious infections and cancer. Some common side effects of rilonacept include, cold symptoms, nausea, stomach pain, diarrhea, numbness or tingly feeling and injection site reaction. For mavrilimumab, there is a theoretical risk for the development of pulmonary alveolar proteinosis, or PAP. PAP is a rare lung disorder in which

surfactant-derived lipoproteins accumulate excessively within pulmonary alveoli due to loss of GM-CSF function. The disease can range in severity from a sub-clinical reduction in diffusion capacity to significant dyspnea during mild exertion. In pre-clinical studies conducted by MedImmune, certain effects were observed in the lungs of non-human primates, which led the FDA to issue a clinical hold with respect to MedImmune's proposed clinical trial in rheumatoid arthritis. Pre-clinical data generated to date suggest mavrilimumab does not reach the lungs in sufficient quantities to induce PAP at clinically relevant doses and human trials thus far have not shown a clinical effect on pulmonary function tests attributable to mavrilimumab. If the results of our trials reveal a high or unacceptable severity and prevalence of these or other side effects, the FDA or applicable foreign regulatory agency may not authorize us to initiate our trials, or if initiated, our clinical trials could be suspended or terminated. The FDA or comparable foreign regulatory authorities could order us to cease further development of, or deny or withdraw approval of, any of our product candidates for any or all targeted indications.

Additionally, if one or more of our product candidates receives marketing approval, and we or others later identify undesirable side effects caused by such product, a number of potentially significant negative consequences could result, including but not limited to:

- regulatory authorities may withdraw approvals of such product and require us to take them off the market;
- regulatory authorities may require the addition of labeling statements, specific warnings, contraindications or field alerts to physicians and pharmacies;
- we may be required to create a REMS plan, which could include a medication guide outlining the risks of such side effects for distribution to patients, a communication plan for healthcare providers and/or other elements to assure safe use;
- we may be required to change the way a product is administered, conduct additional clinical trials or change the labeling of products;
- we may be subject to limitations on how we promote the product, or sales of the product may decrease significantly;
- we could be sued and held liable for harm caused to patients;
- the product may become less competitive; and
- our reputation may suffer.

Any of these events could prevent us from achieving or maintaining market acceptance of the particular product candidate, if approved, and could significantly harm our business, results of operations and prospects.

Prior to our in-license or acquisition of rilonacept, mavrilimumab, KPL-716, KPL-045, and KPL-404, we were not involved in the development of these product candidates and, as a result, we are dependent on Regeneron, MedImmune, Biogen, Novo Nordisk and Primatope having accurately reported the results and correctly collected and interpreted the data from all pre-clinical and clinical trials conducted prior to our acquisition.

We had no involvement with or control over the pre-clinical and clinical development of any of our product candidates prior to our in-license or acquisition of them. We are dependent on Regeneron, MedImmune, Biogen, Novo Nordisk, and Primatope having conducted their research and development in accordance with the applicable protocols and legal, regulatory and scientific standards; having accurately reported the results of all pre-clinical studies and clinical trials conducted prior to our in-license or acquisition; and having correctly collected and interpreted the

data from these trials. If these activities were not compliant, accurate or correct, the clinical development, regulatory approval, or commercialization of one or more of our product candidates will be adversely affected.

If we cannot replicate positive results from earlier pre-clinical studies conducted by us or the companies from whom we have licensed or acquired or may in the future license or acquire our product candidates in our later clinical trials, we may be unable to successfully develop, obtain regulatory approval for and commercialize our product candidates. We have not yet generated any human data demonstrating efficacy in the diseases in which we are studying our product candidates, and we may never be able to do so.

Positive results from our pre-clinical studies, and any positive results we may obtain from our early clinical trials of our product candidates or from the clinical trials conducted by the companies from whom we licensed or acquired or may in the future license or acquire our product candidates, may not necessarily be predictive of the results from our required later pre-clinical studies and clinical trials. Similarly, even if we are able to complete our planned pre-clinical studies or clinical trials of our product candidates, the positive results from the pre-clinical studies and clinical trials of our product candidates may not be replicated in our subsequent pre-clinical studies or clinical trial results. None of our product candidates have been studied for the indications in which we are developing them, and we cannot provide any assurance that their development will be successful. For example, although rilonacept is FDA-approved for the treatment of CAPS, and mavrilimumab has been studied in Phase 2 clinical trials for the treatment of RA, their safety and efficacy has not been determined in recurrent pericarditis or GCA, respectively, and each may fail to receive regulatory approval for those indications.

Many companies in the pharmaceutical and biotechnology industries have suffered significant setbacks in late-stage clinical trials after achieving positive results in early-stage development and we cannot be certain that we will not face similar setbacks. These setbacks have been caused by, among other things, pre-clinical findings made while clinical trials were underway or safety or efficacy observations made in pre-clinical studies and clinical trials, including previously unreported adverse events. Moreover, pre-clinical and clinical data are often susceptible to varying interpretations and analyses and many companies that believed their product candidates performed satisfactorily in pre-clinical studies and clinical trials nonetheless failed to obtain FDA or EMA approval. Furthermore, the approval policies or regulations of the FDA or the applicable foreign regulatory agencies may significantly change in a manner rendering our clinical data insufficient for approval, which may lead to the FDA or any foreign regulatory bodies delaying, limiting or denying approval of our product candidates.

Interim "top-line" and preliminary data from our clinical trials that we announce or publish from time to time may change as more patient data become available and are subject to audit and verification procedures that could result in material changes in the final data.

From time to time, we may publish interim "top-line" or preliminary data from our clinical trials. Preliminary or interim data from clinical trials that we may complete are subject to the risk that one or more of the clinical outcomes may materially change as patient enrollment continues and more patient data become available. Preliminary or interim data also remain subject to audit and verification procedures that may result in the final data being materially different from the preliminary data we previously published. As a result, interim and preliminary data should be viewed with caution until the final data are available. Adverse differences between preliminary or interim data and final data could significantly harm our business prospects.

Regulatory approval processes are lengthy, time consuming and inherently unpredictable. If we are not able to obtain, or if there are delays in obtaining, required regulatory approvals for our product candidates, we will not be able to commercialize, or will be delayed in commercializing, our product candidates and our ability to generate revenue will be materially impaired.

Our product candidates and the activities associated with their development and commercialization, including their design, testing, manufacture, safety, efficacy, recordkeeping, labeling, storage, approval, advertising, promotion, sale, distribution, import and export are subject to comprehensive regulation by the FDA and other regulatory agencies in the United States and by comparable authorities in other countries. Before we can commercialize any of our product candidates, we must obtain marketing approval. We have not received approval or clearance to market any of our product candidates from regulatory authorities in any jurisdiction and it is possible that none of our product candidates or any product candidates we may seek to develop in the future will ever obtain regulatory approval or clearance. We have only limited experience in filing and supporting the applications necessary to gain regulatory approvals and may need to rely on third-party CROs and regulatory consultants to assist us in this process. Securing regulatory approval requires the submission of extensive pre-clinical and clinical data and supporting information to the various regulatory authorities for each therapeutic indication to establish the product candidate's safety and efficacy. Securing regulatory approval also requires the submission of information about the biologic manufacturing process to and inspection of manufacturing facilities by, the relevant regulatory authority. Our product candidates may not be effective, may be only moderately effective or may prove to have undesirable or unintended side effects, toxicities or other characteristics that may preclude our obtaining marketing approval or prevent or limit commercial use. The FDA and other regulatory authorities have substantial discretion in the approval process, and determining when or whether regulatory approval will be obtained for a product candidate. Even if we believe the data collected from clinical trials are promising, such data may not be sufficient to support approval by the FDA or any other regulatory authority.

The process of obtaining regulatory approvals, both in the United States and in other countries, is expensive, may take many years if additional clinical trials are required, if approval is obtained at all, and can vary substantially based upon a variety of factors, including the type, complexity and novelty of the product candidates involved. Changes in marketing approval policies during the development period, changes in or the enactment of additional statutes or regulations, or changes in regulatory review for each submitted BLA, or equivalent application types, may cause delays in the approval or rejection of an application. The FDA and comparable authorities in other countries have substantial discretion in the approval process and may refuse to accept any application or may decide that our data are insufficient for approval and require additional pre-clinical, clinical or other trials. Approval by the FDA in the United States, if obtained, does not ensure approval by regulatory authorities in other countries or jurisdictions. Our product candidates could be delayed in receiving, or fail to receive, regulatory approval for many reasons, including the following:

- the FDA or comparable foreign regulatory authorities may disagree with the number, design or implementation of our clinical trials to support further development or approval;
- we may be unable to demonstrate to the satisfaction of the FDA or comparable foreign regulatory authorities that a product candidate is safe and effective for its proposed indication;
- the results of clinical trials may not meet the level of statistical significance required by the FDA or comparable foreign regulatory authorities for approval;

- we may be unable to demonstrate that a product candidate's clinical and other benefits outweigh its safety risks;
- the FDA or comparable foreign regulatory authorities may disagree with our interpretation of data from pre-clinical studies or clinical trials;
- the FDA or comparable foreign regulatory authorities may disagree that we have provided sufficient safety data or adequately demonstrated clinical benefit in a patient population or subpopulation studied in the clinical trial;
- the data collected from clinical trials of our product candidates may not be sufficient to support the submission of a BLA or other submission or to obtain regulatory approval in the United States or elsewhere;
- the FDA or comparable foreign regulatory authority could require us to collect additional data or conduct additional clinical studies;
- the number of patients required for clinical trials of our product candidates may be larger than we anticipate, enrollment in these clinical trials may be slower than we anticipate, participants may drop out of these clinical trials at a higher rate than we anticipate or we may fail to recruit suitable patients to participate in a trial;
- our third-party contractors may fail to comply with regulatory requirements or meet their contractual obligations to us in a timely manner, or at all;
- the FDA or comparable foreign regulatory authorities may not believe that we have sufficiently demonstrated our ability to manufacture the products to the requisite level of quality standards, or they may fail to approve our manufacturing processes or facilities, or the manufacturing processes or facilities of third-party manufacturers with which we contract for clinical and commercial supplies;
- the supply or quality of our product candidates or other materials necessary to conduct clinical trials of our product candidates may be insufficient or inadequate;
- our product candidates may have undesirable side effects or other unexpected characteristics, causing us or our investigators, regulators or institutional review boards to suspend or terminate the clinical trials; and
- the approval policies or regulations of the FDA or comparable foreign regulatory authorities may significantly change in a manner rendering our clinical data insufficient for approval.

In addition, even if we were to obtain approval for one or more of our product candidates, regulatory authorities may approve any of our product candidates for fewer or more limited indications than we request. For example, in connection with our KPL-716 program, regulatory authorities may recognize a narrower patient population as having prurigo nodularis or define the disease differently than we do. Furthermore, regulatory authorities may not approve the price we intend to charge, may grant approval contingent on the performance of costly post-marketing clinical trials, may impose certain post-marketing requirements that impose limits on our marketing and distribution activities, or may approve a product candidate with a label that does not include the labeling claims necessary or desirable for the successful commercialization of that product candidate. Any of the foregoing scenarios could materially harm the commercial prospects for our product candidates.

If we experience delays in obtaining approval or if we fail to obtain approval of our product candidates, the commercial prospects for our product candidates may be harmed and our ability to generate revenue will be materially impaired.

Our product candidates regulated as biologics in the United States may face competition sooner than anticipated.

In the United States, the Biologics Price Competition and Innovation Act of 2009, or BPCIA, created an abbreviated approval pathway for biological products that are biosimilar to or interchangeable with an FDA-licensed reference biological product. Under the BPCIA, an application for a biosimilar product may not be submitted to the FDA until four years following the date that the reference product was first licensed by the FDA. In addition, the approval of a biosimilar product may not be made effective by the FDA until 12 years from the date on which the reference product was first licensed. During this 12-year period of exclusivity running from this 2008 approval, another company may still market a competing version of the reference product if the FDA approves a full BLA for the competing product containing the sponsor's own pre-clinical data and data from adequate and well-controlled clinical trials to demonstrate the safety, purity and potency of their product. The law is complex and is still being interpreted and implemented by the FDA. As a result, its ultimate impact, implementation and meaning are subject to uncertainty. While it is uncertain when processes intended to implement the BPCIA may be fully adopted by the FDA, any such processes could have a material adverse effect on the future commercial prospects of our product candidates.

Rilonacept was approved as a biological product under a BLA for the treatment of CAPS in 2008, and we believe it should qualify for the 12-year period of exclusivity against any biosimilars. However, there is a risk that this exclusivity could be shortened due to congressional action or otherwise, or that the FDA will not consider rilonacept, or any of our other product candidates, to be reference products for competing products, potentially creating the opportunity for generic competition sooner than anticipated. In addition, we plan to submit a supplemental BLA for rilonacept for the treatment of recurrent pericarditis, and the 12-year exclusivity period does not attach to the approval of a supplemental BLA.

Other aspects of the BPCIA, some of which may impact the BPCIA exclusivity provisions, have also been the subject of recent litigation. Moreover, the extent to which a biosimilar, once approved, will be substituted for a reference product in a way that is similar to traditional generic substitution for non-biological products is not yet clear, and will depend on a number of marketplace and regulatory factors that are still developing.

Even if we obtain marketing approval of our product candidates in a major pharmaceutical market such as the United States or the European Union, we may not obtain approval or commercialize our product candidates in other markets, which would limit our ability to realize their full market potential.

In order to market any products in a country or territory, we must establish and comply with numerous and varying regulatory requirements of such country or territory regarding safety and efficacy. Clinical trials conducted in one country may not be accepted by regulatory authorities in other countries, and regulatory approval in one country does not mean that regulatory approval will be obtained in any other country. Approval procedures vary among countries and can involve additional product testing and validation and additional administrative review periods. Seeking regulatory approvals in all markets may require additional pre-clinical studies or clinical trials, which would be costly and time consuming. Regulatory requirements can vary widely from country to country and could delay or prevent the introduction of our product candidates in those countries. Satisfying these and other regulatory requirements is costly, time consuming, uncertain and subject to unanticipated delays. In addition, our failure to obtain regulatory approval in any country may delay or have negative effects on the process for regulatory approval in other countries.

We may seek orphan drug designation for some of our product candidates and we may be unsuccessful, or may be unable to maintain the benefits associated with orphan drug designation, including the potential for market exclusivity, for any product candidate for which we obtain orphan drug designation.

As part of our business strategy, we intend to seek orphan drug designation for certain of our product candidates, such as rilonacept, and we may be unsuccessful or unable to maintain the associated benefits. Regulatory authorities in some jurisdictions, including the United States and Europe, may designate drugs or biologics intended to treat relatively small patient populations as orphan drug products. Under the U.S. Orphan Drug Act, the FDA may designate a drug or biologic as an orphan drug if it is intended to treat a rare disease or condition, which is generally defined as a patient population of fewer than 200,000 individuals in the United States, or a patient population greater than 200,000 in the United States where there is no reasonable expectation that the cost of developing the drug will be recovered from sales in the United States.

In the European Union, the European Commission grants orphan drug designation after receiving the opinion of the EMA's Committee for Orphan Medicinal Products on an Orphan Drug Designation application. In the European Union, Orphan Drug Designation is intended to promote the development of drugs that are intended for the diagnosis, prevention or treatment of life-threatening or chronically debilitating conditions affecting not more than five in 10,000 persons in the European Union and for which no satisfactory method of diagnosis, prevention, or treatment has been authorized (or the product would be a significant benefit to those affected). Additionally, orphan designation is granted for drugs intended for the diagnosis, prevention or treatment of a life-threatening, seriously debilitating or serious and chronic condition when, without incentives, it is unlikely that sales of the drug in the European Union would be sufficient to justify the necessary investment in developing the drug. In the European Union, Orphan Drug Designation entitles a party to financial incentives such as reduction of fees or fee waivers, as well as potential marketing exclusivity.

In addition, if a drug or biologic with an orphan drug designation subsequently receives the first marketing approval for the indication for which it has such designation, the drug or biologic is entitled to a period of marketing exclusivity, which precludes the FDA from approving another marketing application for the same drug and indication for that time period, except in limited circumstances. If our competitors are able to obtain orphan drug exclusivity prior to use, for products that constitute the "same drug" and treat the same indications as our product candidates, we may not be able to have competing products approved by the applicable regulatory authority for a significant period of time. The applicable period is seven years in the United States and ten years in the European Union. The European Union exclusivity period can be reduced to six years if a drug no longer meets the criteria for orphan drug designation or if the drug is sufficiently profitable so that market exclusivity is no longer justified.

Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent FDA from approving the same or a different drug in another indication. Even after an orphan drug is approved, the FDA can subsequently approve a later application for the same drug for the same condition if the FDA concludes that the later drug is clinically superior in that it is shown to be safer in a substantial portion of the target populations, more effective or makes a major contribution to patient care. In addition, a designated orphan drug may not receive orphan drug exclusivity if it is approved for a use that is broader than the indication for which it received orphan designation. Moreover, orphan drug exclusive marketing rights in the United States may be lost if the FDA later determines that the request for designation was materially defective or if we are unable to manufacture sufficient quantities of the product to meet the needs of patients with the rare disease

or condition. Orphan drug designation neither shortens the development time or regulatory review time of a drug nor gives the drug any advantage in the regulatory review or approval process. While we intend to seek Orphan Drug Designation for our other product candidates in addition to rilonacept, we may never receive such designations. Even if we do receive such designations, there is no guarantee that we will enjoy the benefits of those designations.

We may seek Breakthrough Therapy designation or Fast Track designation by the FDA for one or more of our product candidates, but we may not receive such designation, and even if we do, such designation may not lead to a faster development or regulatory review or approval process and it does not increase the likelihood that our product candidates will receive marketing approval.

We may seek a Breakthrough Therapy or Fast Track designation for some of our product candidates. A breakthrough therapy is defined as a drug that is intended, alone or in combination with one or more other drugs or biologics, to treat a serious or life-threatening disease or condition and preliminary clinical evidence indicates that the drug may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. For drugs or biologics that have been designated as breakthrough therapies, interaction and communication between the FDA and the sponsor of the trial can help to identify the most efficient path for clinical development while minimizing the number of patients placed in ineffective control regimens. Drugs or biologics designated as breakthrough therapies by the FDA may also be eligible for accelerated approval.

If a product candidate is intended for the treatment of a serious or life-threatening condition and clinical or pre-clinical data demonstrate the potential to address unmet medical needs for this condition, the sponsor may apply for Fast Track Designation. The FDA has broad discretion whether or not to grant this designation, so even if we believe a particular product candidate is eligible for this designation, we cannot assure you that the FDA would decide to grant it. Even if we have obtained Fast Track Designation for one or more of our product candidates, we may not experience a faster development process, review or approval compared to non-expedited FDA review procedures. In addition, the FDA may withdraw Fast Track Designation for any product candidate that is granted if it believes that the designation is no longer supported. Fast Track Designation alone does not guarantee qualification for the FDA's priority review procedures.

Whether to grant Breakthrough Therapy or Fast Track Designation is within the discretion of the FDA. Accordingly, even if we believe one of our product candidates meets the criteria for these designations, the FDA may disagree and instead determine not to make such designation. In any event, the receipt of either of these designations for a product candidate may not result in a faster development process, review or approval compared to product candidates considered for approval under non-expedited FDA review procedures and does not assure ultimate approval by the FDA. In addition, even if one or more of our product candidates qualify for either of these designations, the FDA may later decide that the product candidates no longer meet the conditions for qualification.

We have never completed a Phase 3 clinical trial or obtained marketing approval for any product candidate and we may be unable to successfully do so for any of our product candidates. Failure to successfully complete any of these activities in a timely manner for any of our product candidates could have a material adverse impact on our business and financial performance.

Conducting a pivotal clinical trial and preparing, and obtaining marketing approval for, a product candidate is a complicated process. Although members of our management team have participated in pivotal trials and obtained marketing approvals for product candidates in the past while employed at other companies, we as a company have not done so. As a result, such

activities may require more time and cost more than we anticipate. Failure to successfully complete, or delays in, any of our eventual pivotal trials or related regulatory submissions would prevent us from or delay us in obtaining regulatory approval for, or clearance of, our product candidates. In addition, it is possible that the FDA may refuse to accept for substantive review any BLA submissions that we submit for our product candidates or may conclude after review of our applications that they are insufficient to obtain marketing approval or clearance of our product candidates. If the FDA does not accept our applications or issue marketing authorizations for our product candidates, it may require that we conduct additional clinical, pre-clinical or manufacturing validation trials and submit that data before it will reconsider our applications. Depending on the extent of these or any other FDA-required trials, approval of any BLA or receipt of other marketing authorizations for any other applications that we submit may be delayed by several years, or may require us to expend more resources than we have available. It is also possible that additional trials, if performed and completed, may not be considered sufficient by the FDA to approve our BLAs or grant other marketing authorizations. Any delay in obtaining, or an inability to obtain, marketing approvals would prevent us from commercializing our product candidates, generating revenues and achieving and sustaining profitability. If any of these outcomes occur, we may be forced to abandon our development efforts for our product candidates, which could significantly harm our business.

Risks Related to Manufacturing and Our Dependence on Third Parties

We contract with third parties for manufacturing our product candidates and for pre-clinical and clinical development and expect to continue to do so for our commercial supply. This reliance on third parties increases the risk that we may not have sufficient quantities of our product candidates or such quantities at an acceptable cost, which could delay, prevent or impair our development or commercialization efforts.

We do not currently own or operate any manufacturing facilities. Although we may build small scale manufacturing facilities for the production of drug substance to support our clinical trials, we rely, and expect to continue to rely, on third parties for the manufacture of our product candidates for the majority of our pre-clinical development and clinical testing, as well as for the commercial manufacture of our product candidates, if approved. We rely on these third parties to develop the processes necessary to produce our product candidates at sufficient quality and quantity to support our development and commercialization efforts. Our reliance increases the risk that we will have insufficient quantities of our product candidates or that our product candidates are not produced at an acceptable cost or quality, which could delay, prevent or impair our development or commercialization efforts.

We plan to enter into agreements with CMOs to produce mavrilimumab beyond our current inventory. We will need to transfer the technology to manufacture mavrilimumab to these CMOs, and these CMOs may decide or be required to adopt different manufacturing protocols or processes. In addition, we will need to produce mavrilimumab using different media and feed compared to the processes that were used by MedImmune to develop our existing inventory. We cannot provide any assurance that the technology transfer or process development will be successful, or that any CMO will be successful in producing mavrilimumab in sufficient quantities or of acceptable quality, if at all. We also contract with Regeneron to produce rilonacept, with CMOs for the manufacture of KPL-716 drug substance and drug product, and CMOs to produce our pre-clinical product candidates, KPL-045 and KPL-404.

The facilities used by our contract manufacturers to manufacture our product candidates may be inspected by the FDA and other comparable regulatory authorities in connection with the submission of our marketing applications to, and review by, the FDA. While we provide oversight of manufacturing activities, we do not and will not control the manufacturing process of, and will be completely dependent on, our contract manufacturers for compliance with cGMPs and other

regulatory requirements in connection with the manufacture of our product candidates. If our contract manufacturers cannot successfully manufacture material that conforms to our specifications and the strict regulatory requirements of the FDA or others, they will not be able to secure and/or maintain regulatory approval for their manufacturing facilities. In addition, we have no control over the ability of our contract manufacturers to maintain adequate quality control, quality assurance and qualified personnel. If the FDA or a comparable foreign regulatory authority does not approve these facilities for the manufacture of our product candidates or if it withdraws any such approval in the future, we may need to find alternative manufacturing facilities, which would significantly impact our ability to develop, obtain regulatory approval for or market our product candidates, if approved. Further, our failure, or the failure of our third-party manufacturers, to comply with applicable regulations could result in sanctions being imposed on us, including clinical holds, fines, injunctions, civil penalties, delays, suspension or withdrawal of approvals, license revocation, seizures or recalls of product candidates, if approved, operating restrictions and criminal prosecutions, any of which could significantly and adversely affect our business and supplies of our product candidates.

Although we have entered into certain agreements for the manufacture of clinical material for our product candidates, we may be unable to establish new agreements on acceptable terms, if at all, with third-party manufacturers for those product candidates. Even if we are able to establish agreements with third-party manufacturers, reliance on third-party manufacturers entails additional risks, including:

- reliance on the third-party for regulatory compliance and quality assurance;
- the possible breach of the manufacturing agreement by the third-party;
- the possible misappropriation of our proprietary information, including our trade secrets and know-how; and
- the possible termination or nonrenewal of the agreement by the third-party at a time that is costly or inconvenient for us.

Our product candidates may compete with other product candidates and approved products for access to manufacturing facilities. There are a limited number of manufacturers that operate under cGMP regulations and that might be capable of manufacturing for us. Further, Regeneron has an exclusive right to produce rilonacept for a period of time.

Any performance failure on the part of our existing or future manufacturers could delay clinical development or marketing approval. If our current CMOs cannot perform as agreed, we may be required to replace such manufacturers. Although we believe that there are several potential alternative manufacturers who could manufacture our product candidates, we may incur added costs and delays in identifying and qualifying any such replacement.

Finding new CMOs or third-party suppliers involves additional cost and requires our management's time and focus. In addition, there is typically a transition period when a new CMO commences work. Although we generally do not begin a clinical trial unless we believe we have on hand, or will be able to obtain, a sufficient supply of our product candidates to complete the clinical trial, any significant delay in the supply of our product candidates or the raw materials needed to produce our product candidates, could considerably delay conducting our clinical trials and potential regulatory approval of our product candidates.

Our current and anticipated future dependence upon others for the manufacture of our product candidates may adversely affect our future profit margins and our ability to commercialize any products that receive marketing approval on a timely and competitive basis.

Our business involves the use of hazardous materials and we and our third-party manufacturers and suppliers must comply with environmental laws and regulations, which can be expensive and restrict how we do business.

Our research and development activities and our third-party manufacturers' and suppliers' activities involve the controlled storage, use and disposal of hazardous materials owned by us, including the components of our product and product candidates and other hazardous compounds. We and our manufacturers and suppliers are subject to laws and regulations governing the use, manufacture, storage, handling and disposal of these hazardous materials. In some cases, these hazardous materials and various wastes resulting from their use are stored at our and our manufacturers' facilities pending their use and disposal. We cannot eliminate the risk of contamination, which could cause an interruption of our commercialization efforts, research and development efforts and business operations, environmental damage resulting in costly clean-up and liabilities under applicable laws and regulations governing the use, storage, handling and disposal of these materials and specified waste products. Although we believe that the safety procedures utilized by our third-party manufacturers for handling and disposing of these materials generally comply with the standards prescribed by these laws and regulations, we cannot guarantee that this is the case or eliminate the risk of accidental contamination or injury from these materials. In such an event, we may be held liable for any resulting damages and such liability could exceed our resources and state or federal or other applicable authorities may curtail our use of certain materials and/or interrupt our business operations. Furthermore, environmental laws and regulations are complex, change frequently and have tended to become more stringent. We cannot predict the impact of such changes and cannot be certain of our future compliance. We do not currently carry biological or hazardous waste insurance coverage.

Manufacturing issues at the facilities of our third-party service providers could cause product shortages, disrupt or delay our clinical trials or regulatory approvals, delay or stop commercialization of our products, and adversely affect our business.

The manufacture of our product candidates is highly regulated, complex and difficult, requiring a multi-step and controlled process, and even minor problems or deviations could result in defects or failures, such as defective products or manufacturing failures. We have limited experience overseeing the manufacturing process of KPL-716 and no experience overseeing the manufacturing process of rilonacept, mavrilimumab, KPL-404 and KPL-045. Due to the highly technical requirements of manufacturing our products and the strict quality and control specifications, we and our third-party providers may be unable to manufacture or supply our product candidates despite our and their efforts. Failure to produce sufficient quantities of our product candidates could delay their development, result in supply shortages for our patients, result in lost revenue, diminish our potential profitability, any of which may lead to lawsuits or could accelerate introduction of competing products to the market.

The manufacture of our product candidates is at high risk of product loss due to contamination, equipment malfunctions, human error or raw material shortages. Deviations from established manufacturing processes could result in reduced production yields, product defects and other supply disruptions. If microbial, viral or other contaminations are discovered in our product candidates or manufacturing facilities, any related production lot could be lost and the relevant manufacturing facilities may need to close for an extended period of time to investigate and remediate the contaminant. Many additional factors could cause production interruptions at our facilities or at the facilities of our third-party providers, including natural disasters, accidents, labor disputes, acts of terrorism or war. The occurrence of any such event could adversely affect our ability to satisfy the required supply for any of our product candidates, successfully complete

pre-clinical and clinical development which would result in additional costs to us or impair our ability to generate revenue and harm our business, financial condition and prospects significantly.

Our third-party providers are required to maintain compliance with cGMP and other stringent requirements and are subject to inspections by the FDA and comparable agencies in other jurisdictions to confirm such compliance. Any delay, interruption or other issues that arise in the manufacture, fill-finish, packaging or storage of our product candidates as a result of a failure of the facilities or operations of third parties to pass any regulatory agency inspection could significantly impair our ability to supply our products and product candidates. Significant noncompliance could also result in the imposition of monetary penalties or other civil or criminal sanctions and damage our reputation.

Any adverse developments affecting the operations of our third-party providers could result in a product shortage of clinical or commercial requirements, withdrawal of our product candidates or any approved products, shipment delays, lot failures or recalls. We may also have to write-off inventory and incur other charges and expenses for products that fail to meet specifications, undertake costly remediation efforts or seek more costly manufacturing alternatives. Such manufacturing issues could increase our cost of goods, cause us to lose potential revenue, reduce our potential profitability or damage our reputation.

The third parties upon whom we rely for the supply of the active pharmaceutical ingredient, drug product and drug substance used in our product candidates are our sole source of supply, and the loss of any of these suppliers could significantly harm our business.

The active pharmaceutical ingredients, or API, drug product and drug substance used in rilonacept, mavrilimumab and KPL-716 are supplied to us from single-source suppliers. For example, although Regeneron has been producing rilonacept for over 10 years, they have a contractual right to be our sole source manufacturer of the product, unless they have a persistent failure to satisfy our supply needs. Our ability to successfully develop our product candidates, and to ultimately supply our commercial products in quantities sufficient to meet the market demand, depends in part on our ability to obtain the API, drug product and drug substance for these product candidates in accordance with regulatory requirements and in sufficient quantities for commercialization and clinical testing. We do not currently have arrangements in place for a redundant or second-source supply of any such API, drug product or drug substance in the event any of our current suppliers of such API, drug product and drug substance cease their operations or stop offering us sufficient quantities of these materials for any reason.

We are not certain that our single-source suppliers will be able to meet our demand for their products, either because of the nature of our agreements with those suppliers, our limited experience with those suppliers or our relative importance as a customer to those suppliers. It may be difficult for us to assess their ability to timely meet our demand in the future based on past performance. While our suppliers have generally met our demand on a timely basis in the past, they may subordinate our needs in the future to their other customers.

In addition, to manufacture rilonacept, mavrilimumab and KPL-716 in the quantities that we believe would be required to meet anticipated market demand, our third-party manufacturers may need to increase manufacturing capacity and, in some cases, we could secure alternative sources of commercial supply, which could involve significant challenges and may require additional regulatory approvals. In addition, the development of commercial-scale manufacturing capabilities may require us and our third-party manufacturers to invest substantial additional funds and hire and retain the technical personnel who have the necessary manufacturing experience. Neither we nor our third-party manufacturers may successfully complete any required increase to existing manufacturing capacity in a timely manner, or at all.

Moreover, if there is a disruption to one or more of our third-party manufacturers' or suppliers' relevant operations the supply of rilonacept, mavrilimumab and KPL-716 will be delayed until such manufacturer or supplier restores the affected facilities or we or they procure alternative manufacturing facilities or sources of supply. Our ability to progress our pre-clinical and clinical programs could be materially and adversely impacted if any of the third-party suppliers upon which we rely were to experience a significant business challenge, disruption or failure due to issues such as financial difficulties or bankruptcy, issues relating to other customers such as regulatory or quality compliance issues, or other financial, legal, regulatory or reputational issues. Additionally, any damage to or destruction of our third-party manufacturers' or suppliers' facilities or equipment may significantly impair our ability to manufacture our product candidates on a timely basis.

Establishing additional or replacement suppliers for the API, drug product and drug substance used in our product candidates, if required, may not be accomplished quickly and can take several years, if at all. Furthermore, despite our efforts, we may be unable to procure a replacement supplier or do so on commercially reasonable terms, which could have a material adverse impact upon our business. If we are able to find a replacement supplier, such replacement supplier would need to be qualified and may require additional regulatory approval, which could result in further delay. While we seek to maintain adequate inventory of the API, drug product and drug substance used in our product candidates, any interruption or delay in the supply of components or materials, or our inability to obtain such API, drug product and drug substance from alternate sources at acceptable prices in a timely manner could impede, delay, limit or prevent our development efforts, which could harm our business, results of operations, financial condition and prospects.

Certain of the raw materials required in the manufacture and the formulation of our product candidates are derived from biological sources. Such raw materials are difficult to procure and may be subject to contamination or recall. Access to and supply of sufficient quantities of raw materials which meet the technical specifications for the production process is challenging, and often limited to single-source suppliers. Finding an alternative supplier could take a significant amount of time and involve significant expense due to the nature of the products and the need to obtain regulatory approvals. If we or our manufacturers are unable to purchase the raw materials necessary for the manufacture of our product candidates on acceptable terms in a timely manner, at sufficient quality levels, or in adequate quantities, if at all, our ability to produce sufficient quantities of our products for clinical or commercial requirements would be negatively impacted. A material shortage, contamination, recall or restriction on the use of certain biologically derived substances or any raw material used in the manufacture of our products could adversely impact or disrupt manufacturing, which would impair our ability to generate revenues from the sale of such product candidates, if approved or cleared.

We rely, and expect to continue to rely, on third parties, including independent investigators and CROs, to conduct our research, pre-clinical studies, clinical trials and other trials for our product candidates. If these third parties do not successfully carry out their contractual duties, comply with regulatory requirements or meet expected deadlines, we may not be able to obtain regulatory approval for or commercialize our product candidates and our business could be substantially harmed.

We do not have the ability to independently conduct pre-clinical studies or clinical trials that comply with the GLPs or GCP requirements, respectively. We rely on medical institutions, clinical investigators, contract laboratories and other third parties, such as CROs, to conduct or otherwise support our GLP-compliant pre-clinical studies and GCP-compliant clinical trials for our product candidates properly and on time. We also rely on third parties to conduct other research related to our product candidates. We expect to rely heavily on these parties for execution of clinical trials for our product candidates and control only certain aspects of their activities. While we have

agreements governing their activities, we control only certain aspects of their activities and have limited influence over their actual performance. The third parties with whom we contract for execution of our GLP-compliant pre-clinical studies and our GCP-compliant clinical trials play a significant role in the conduct of these trials and trials and the subsequent collection and analysis of data. These third parties are not our employees and, except for restrictions imposed by our contracts with such third parties, we have limited ability to control the amount or timing of resources that they devote to our programs. Although we rely on these third parties to conduct our GLP-compliant pre-clinical studies and GCP-compliant clinical trials, we will remain responsible for ensuring that each of our clinical trials is conducted in accordance with the applicable protocol, legal and regulatory requirements and scientific standards, and our reliance on these third parties does not and will not relieve us of our regulatory responsibilities. For any violations of laws and regulations during the conduct of our clinical trials, we could be subject to warning letters or enforcement action that may include civil penalties up to and including criminal prosecution.

We and our CROs will be required to comply with regulations, including GCPs, for conducting, monitoring, recording and reporting the results of clinical trials to ensure that the data and results are scientifically credible and accurate, and that the trial patients are adequately informed of the potential risks of participating in clinical trials and their rights are protected. These regulations are enforced by the FDA, the Competent Authorities of the Member States of the European Economic Area and comparable foreign regulatory authorities for any products in clinical development. The FDA enforces GCP regulations through periodic inspections of clinical trial sponsors, principal investigators and trial sites. If we or our CROs fail to comply with applicable GCPs, the clinical data generated in our clinical trials may be deemed unreliable and the FDA or comparable foreign regulatory authorities may require us to perform additional clinical trials before approving our marketing applications. We cannot ensure that, upon inspection, the FDA will determine that any of our future clinical trials will comply with GCPs. In addition, our clinical trials must be conducted with product candidates produced under cGMPs regulations. Our failure or the failure of our CROs to comply with these regulations may require us to repeat clinical trials, which would delay the regulatory approval process and could also subject us to enforcement action. We also are required to register certain clinical trials and post the results of completed clinical trials on a government-sponsored database, ClinicalTrials.gov, within certain timeframes. Failure to do so when required can result in fines, adverse publicity and civil and criminal sanctions.

Although we intend to design the clinical trials for our product candidates, CROs will conduct all of the clinical trials. As a result, many important aspects of our development programs, including their conduct and timing, will be outside of our direct control. Our reliance on third parties to conduct future clinical trials will also result in less direct control over the management of data developed through clinical trials than would be the case if we were relying entirely upon our own staff. Communicating with outside parties can also be challenging, potentially leading to mistakes as well as difficulties in coordinating activities. Outside parties may:

- have staffing difficulties;
- fail to comply with contractual obligations;
- experience regulatory compliance issues;
- undergo changes in priorities or become financially distressed; or
- form relationships with other entities, some of which may be our competitors.

These factors may materially adversely affect the willingness or ability of third parties to conduct our clinical trials and may subject us to unexpected cost increases that are beyond our control. If the CROs do not perform clinical trials in a satisfactory manner, breach their obligations to us or fail to comply with regulatory requirements, the development, regulatory approval and

commercialization of our product candidates may be delayed, we may not be able to obtain regulatory approval and commercialize our product candidates, or our development program materially and irreversibly harmed. If we are unable to rely on clinical data collected by our CROs, we could be required to repeat, extend the duration of or increase the size of any clinical trials we conduct and this could significantly delay commercialization and require significantly greater expenditures.

These third parties are not our employees and we are not able to control, other than by contract, the amount of resources, including time, which they devote to our clinical trials. If our independent investigators or CROs fail to devote sufficient resources to the development of our product candidates, or if their performance is substandard, it may delay or compromise the prospects for approval and commercialization of our product candidates. In addition, the use of third-party service providers requires us to disclose our proprietary information to these parties, which could increase the risk that this information is misappropriated.

If the third parties conducting our pre-clinical studies or our clinical trials do not perform their contractual duties or obligations, experience significant business challenges, disruptions or failures, do not meet expected deadlines, terminate their agreements with us or need to be replaced, or if the quality or accuracy of the data they obtain is compromised due to their failure to adhere to our protocols or to GCPs, or for any other reason, we may need to enter into new arrangements with alternative third parties. This could be difficult, costly or impossible, and our pre-clinical studies or clinical trials may need to be extended, delayed, terminated or repeated. As a result, we may not be able to obtain regulatory approval in a timely fashion, or at all, for the applicable product candidate, our financial results and the commercial prospects for our product candidates would be harmed, our costs could increase, and our ability to generate revenues could be delayed.

If any of our relationships with these third-party CROs terminate, we may not be able to enter into arrangements with alternative third-party service providers, at all or on commercially reasonable terms. If CROs do not successfully carry out their contractual duties or obligations or meet expected deadlines, if they need to be replaced or if the quality or accuracy of the clinical data they obtain is compromised due to the failure to adhere to our clinical protocols, regulatory requirements or for other reasons, any clinical trials such CROs are associated with may be extended, delayed or terminated, and we may not be able to obtain regulatory approval for or successfully commercialize our product candidates. As a result, we believe that our financial results and the commercial prospects for our product candidates in the subject indication would be harmed, our costs could increase and our ability to generate revenue could be delayed.

Furthermore, principal investigators for our clinical trials may serve as scientific advisors or consultants to us from time to time and may receive cash or equity compensation in connection with such services. If these relationships and any related compensation result in perceived or actual conflicts of interest, or a regulatory authority concludes that the financial relationship may have affected the interpretation of the trial, the integrity of the data generated at the applicable clinical trial site may be questioned and the utility of the clinical trial itself may be jeopardized, which could result in the delay or rejection of the marketing application we submit. Any such delay or rejection could prevent or delay us from commercializing our current or future product candidates.

Any collaboration arrangements that we may enter into in the future may not be successful, which could adversely affect our ability to develop and commercialize our product candidates.

We may seek collaboration arrangements for the commercialization, or potentially for the development, of certain of our product candidates depending on the merits of retaining commercialization rights for ourselves as compared to entering into collaboration arrangements. We will face, to the extent that we decide to enter into collaboration agreements, significant competition

in seeking appropriate collaborators. Moreover, collaboration arrangements are complex and time-consuming to negotiate, document, implement and maintain. We may not be successful in our efforts to establish and implement collaborations or other alternative arrangements should we so chose to enter into such arrangements. The terms of any collaborations or other arrangements that we may establish may not be favorable to us. In addition, our right to grant a sublicense of intellectual property licensed to us under certain of our current agreements requires the consent of the applicable licensor.

Any future collaborations that we enter into may not be successful. The success of our collaboration arrangements will depend heavily on the efforts and activities of our collaborators. Collaborations are subject to numerous risks, which may include risks that:

- collaborators have significant discretion in determining the efforts and resources that they will apply to collaborations;
- a collaborator with marketing, manufacturing and distribution rights to one or more products may not commit sufficient resources to or otherwise not perform satisfactorily in carrying out these activities;
- collaborators may not properly maintain or defend our intellectual property rights or may use our intellectual property or proprietary information in a way that gives rise to actual or threatened litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential liability;
- collaborations may be terminated, and, if terminated, this may result in a need for additional capital to pursue further development or commercialization of the applicable current or future product candidates;
- collaborators may own or co-own intellectual property covering products that results from our collaborating with them, and in such cases, we would not have the exclusive right to develop or commercialize such intellectual property;
- disputes may arise with respect to the ownership of any intellectual property developed pursuant to our collaborations; and
- a collaborator's sales and marketing activities or other operations may not be in compliance with applicable laws resulting in civil or criminal proceedings.

Our reliance on third parties requires us to share our trade secrets, which increases the possibility that a competitor will discover them or that our trade secrets will be misappropriated or disclosed.

Because we rely on third parties to develop and manufacture our product candidates, we must, at times, share trade secrets with them. We seek to protect our proprietary technology in part by entering into confidentiality agreements and, if applicable, material transfer agreements, collaborative research agreements, consulting agreements or other similar agreements with our collaborators, advisors, employees and consultants prior to beginning research or disclosing proprietary information. These agreements typically limit the rights of the third parties to use or disclose our confidential information, such as trade secrets. Despite the contractual provisions employed when working with third parties, the need to share trade secrets and other confidential information increases the risk that such trade secrets become known by our competitors, are inadvertently incorporated into the technology of others, or are disclosed or used in violation of these agreements. Given that our proprietary position is based, in part, on our know-how and trade secrets, a competitor's discovery of our trade secrets or other unauthorized use or disclosure would impair our competitive position and may harm our business. To the extent that we share trade secrets of third parties that are licensed to us, unauthorized use or disclosure could expose us to liability.

Risks Related to Competition, Retaining Key Employees and Managing Growth

We face substantial competition, which may result in others discovering, developing or commercializing drugs before or more successfully than we do.

The development and commercialization of new drugs and biologics is highly competitive. We face competition with respect to our current product candidates, and will face competition with respect to any product candidates that we may seek to develop or commercialize in the future, from major pharmaceutical companies, specialty pharmaceutical companies and biotechnology companies worldwide. There are a number of large pharmaceutical and biotechnology companies that currently market and sell drugs or biologics are pursuing the development of therapies in the fields in which we are interested. Some of these competitive products and therapies are based on scientific approaches that are the same as or similar to our approach, and others are based on entirely different approaches. Potential competitors also include academic institutions, government agencies and other public and private research organizations that conduct research, seek patent protection and establish collaborative arrangements for research, development, manufacturing and commercialization.

While we are not aware of any therapies currently approved or actively continuing clinical trials in recurrent pericarditis, there is one currently marketed product that modulates the signaling of IL-1a and IL-1b, anakinra (KINERET), and one currently marketed product that modulates the signaling of IL-1b, canakinumab (ILARIS). There are other therapies which modulate IL-1a and IL-1b in various stages of clinical development for diseases other than recurrent pericarditis from companies that include Abbvie, Inc., XBiotech Inc. and Handok Inc. We expect mavrilimumab, if approved, to experience competitive pressure from tocilizumab (ACTEMRA), which was approved in 2017 for use in GCA in combination with glucocorticoids. Additional competition may be experienced from upadacitinib from AbbVie, which is expected to enter clinical trials in GCA in 2018. In addition, Eli Lilly is conducting clinical trials in GCA for baricitinib, and Sanofi S.A.,/ Regeneron intend to initiate a Phase 3 clinical trial in GCA for sarilumab (KEVZARA) in 2018. KPL-716, if approved for atopic dermatitis, will face competitive pressure from dupilumab (DUPIXENT), which is approved to treat atopic dermatitis. KPL-716 may face additional competition from several systemically administered products currently in development for atopic dermatitis including upadacitinib, PF-04965842, ANB-020, nemolizumab, baricitinib, ASn002, GBR-830, ZPL-389, PF-06817024, MEDI9314, tralokinumab and lebrikizumab. Multiple therapies are in development for prurigo nodularis and any that receive FDA approval for this indication will be likely competitors to KPL-716. These products include nemolizumab, serlopitant and nalbuphine ER.

Many of the companies against which we are competing or against which we may compete in the future have significantly greater financial resources and expertise in research and development, manufacturing, pre-clinical testing, conducting clinical trials, obtaining regulatory approvals and marketing approved products than we do. Mergers and acquisitions in the pharmaceutical, biotechnology and diagnostic industries may result in even more resources being concentrated among a smaller number of our competitors. Smaller or early stage companies may also prove to be significant competitors, particularly through collaborative arrangements with large and established companies. These competitors also compete with us in recruiting and retaining qualified scientific and management personnel and establishing clinical trial sites and patient registration for clinical trials, as well as in acquiring technologies complementary to, or necessary for, our programs.

Our commercial opportunity could be reduced or eliminated if our competitors develop and commercialize products that are safer, more effective, have fewer or less severe side effects, are more convenient or are less expensive than any drugs that we or our collaborators may develop. Our competitors also may obtain FDA or other regulatory approval for their products more rapidly

than we may obtain approval for ours, which could result in our competitors establishing a strong market position before we or our collaborators are able to enter the market. The key competitive factors affecting the success of all of our product candidates, if approved, are likely to be their efficacy, safety, convenience, price, the effectiveness of companion diagnostics in guiding the use of related products, market acceptance by physicians and patients, the level of generic competition and the availability of reimbursement from government and other third-party payors.

Our future success depends on our ability to retain key executives and to attract, retain and motivate qualified personnel.

We have a limited operating history and are highly dependent on the research and development, clinical, commercial and business development expertise of Sanj K. Patel, our Chairman and Chief Executive Officer, Stephen Mahoney, our President and Chief Operating Officer, and John F. Paolini, M.D., Ph.D., our Chief Medical Officer, as well as the other principal members of our management, scientific and clinical team. Although we have entered into employment agreements with our executive officers, each of them may terminate their employment with us at any time. We do not maintain "key person" insurance for any of our executives or other employees. In addition, we rely on consultants and advisors, including scientific and clinical advisors, to assist us in formulating our research and development and commercialization strategy. Our consultants and advisors may be employed by employers other than us and may have commitments under consulting or advisory contracts with other entities that may limit their availability to us. If we are unable to continue to attract and retain high quality personnel, our ability to pursue our growth strategy will be limited.

Recruiting and retaining qualified scientific, clinical, manufacturing and sales and marketing personnel will also be critical to our success. The failure to recruit, or the loss of the services of our executive officers or other key employees could impede the achievement of our research, development and commercialization objectives and seriously harm our ability to successfully implement our business strategy. Furthermore, replacing executive officers and key employees may be difficult and may take an extended period of time because of the limited number of individuals in our industry with the breadth of skills and experience required to successfully develop, gain regulatory approval of and commercialize products. Competition to hire from this limited pool is intense, and we may be unable to hire, train, retain or motivate these key personnel on acceptable terms given the competition among numerous pharmaceutical and biotechnology companies for similar personnel. We also experience competition for the hiring of scientific and clinical personnel from universities and research institutions. Failure to succeed in clinical trials may make it more challenging to recruit and retain qualified scientific personnel. If we are not able to continue to attract and retain, on acceptable terms, the qualified personnel necessary for the continued development of our business, we may not be able to sustain our operations or growth.

We will need to develop and expand our company, and we may encounter difficulties in managing this development and expansion, which could disrupt our operations.

In connection with becoming a public company, we expect to increase our number of employees and the scope of our operations. To manage our anticipated development and expansion, we must continue to implement and improve our managerial, operational and financial systems, expand our facilities and continue to recruit and train additional qualified personnel. Also, our management may need to divert a disproportionate amount of its attention away from its day-to-day activities and devote a substantial amount of time to managing these development activities. Due to our limited resources, certain employees may need to perform activities that are beyond their regular scope of work, and we may not be able to effectively manage the expansion of our operations or recruit and train additional qualified personnel. This may result in weaknesses in

our infrastructure, give rise to operational mistakes, loss of business opportunities, loss of employees and reduced productivity among remaining employees. The physical expansion of our operations may lead to significant costs and may divert financial resources from other projects, such as the development of our product candidates. If our management is unable to effectively manage our expected development and expansion, our expenses may increase more than expected, our ability to generate revenue could be reduced and we may not be able to implement our business strategy. Our future financial performance and our ability to commercialize our product candidates, if approved, and compete effectively will depend, in part, on our ability to effectively manage the future development and expansion of our company.

We may not be successful in executing our growth strategy to identify, discover, develop, in-license or acquire additional product candidates or our growth strategy may not deliver the anticipated results.

We plan to source new product candidates that are complementary to our existing product candidates through our internal discovery program, or in-licensing or acquiring them from other companies or academic institutions. If we are unable to identify, discover, develop, in-license or acquire and integrate product candidates in accordance with this strategy, our ability to pursue this part of our growth strategy would be limited.

Research programs and business development efforts to identify new product candidates require substantial technical, financial and human resources. We may focus our efforts and resources on potential programs or product candidates that ultimately prove to be unsuccessful. In-licensing and acquisitions of technology often require significant payments, expenses and will consume additional resources. We will need to devote a substantial amount of time and personnel to research, develop and commercialize any acquired technology, in addition to our existing portfolio of programs. Our research programs, business development efforts or licensing attempts may fail to yield additional complementary or successful product candidates for clinical development and commercialization for a number of reasons, including, but not limited to, the following:

- our research or business development methodology or search criteria and process may be unsuccessful in identifying potential product candidates with a high probability of success for development progression;
- we may not be able or willing to assemble sufficient resources or expertise to in-license, acquire or discover additional product candidates;
- for product candidates we seek to in-license or acquire, we may not be able to agree to acceptable terms with the licensor or owner of those product candidates;
- our product candidates may not succeed in pre-clinical studies or clinical trials;
- we may not succeed in formulation or process development;
- our product candidates may be shown to have harmful side effects or may have other characteristics that may make the products unmarketable or unlikely to receive regulatory approval;
- competitors may develop alternatives that render our product candidates obsolete or less attractive;
- product candidates that we develop may be covered by third parties' patents or other exclusive rights;

- product candidates that we develop may not allow us to leverage our expertise and our development and commercial infrastructure as currently expected;
- the market for a product candidate may change during our program so that such a product may become unreasonable to continue to develop;
- a product candidate may not be capable of being produced in commercial quantities at an acceptable cost, or at all; and
- a product candidate may not be accepted as safe and effective by patients, the medical community or third-party payors.

If any of these events occurs, we may not be successful in executing our growth strategy or our growth strategy may not deliver the anticipated results.

Risks Related to Intellectual Property

If we are unable to adequately protect our proprietary technology or obtain and maintain patent protection for our technology and products, if the scope of the patent protection obtained is not sufficiently broad, or if the terms of our patents are insufficient to protect our product candidates for an adequate amount of time, our competitors could develop and commercialize technology and products similar or identical to ours, and our ability to successfully commercialize our technology and products may be materially impaired.

Our commercial success depends in part on our ability to obtain and maintain proprietary or intellectual property protection in the United States and other countries for our product candidates, including rilonacept, mavrilimumab and KPL-716. We seek to protect our proprietary and intellectual property position by, among other methods, filing patent applications in the United States and abroad related to our proprietary technology, inventions and improvements that are important to the development and implementation of our business. We also rely on trade secrets, know-how and continuing technological innovation to develop and maintain our proprietary and intellectual property position.

We acquire, in-license and file patent applications directed to our product candidates in an effort to establish intellectual property positions directed to their compositions of matter as well as uses of these product candidates in the treatment of diseases. Our intellectual property includes patents and patent applications that we own as well as patents and patent applications that we in-license. For example, we have a field-specific exclusive license under a license agreement with Regeneron, or the Regeneron Agreement, to patent applications and patents relating to rilonacept, an exclusive license under a license agreement with MedImmune, or the MedImmune Agreement, to patent applications and patents relating to mavrilimumab, and an exclusive license under our license agreement with Novo Nordisk, or the Novo Nordisk Agreement, to patent applications and patents relating to KPL-045.

Certain provisions in our intellectual property agreements may be susceptible to multiple interpretations. The resolution of any contract interpretation disagreement that may arise could affect the scope of our rights to the relevant intellectual property or technology, or affect financial or other obligations under the relevant agreement, either of which could have a material adverse effect on our business, financial condition, results of operations and prospects.

We or our licensors have not pursued or maintained, and may not pursue or maintain in the future, patent protection for our products in every country or territory in which we may sell our products. In addition, we cannot be sure that any of our pending patent applications or pending trademark applications will issue or that, if issued, they have or will issue in a form that will be advantageous to us. The United States Patent and Trademark Office, or the USPTO, international

patent offices or judicial bodies may deny or significantly narrow claims made under our patent applications and our issued patents may be successfully challenged, may be designed around, or may otherwise be of insufficient scope to provide us with protection for our commercial products. Further, the USPTO, international trademark offices or judicial bodies may deny our trademark applications and, even if published or registered, these trademarks may not effectively protect our brand and goodwill. Like patents, trademarks also may be successfully opposed or challenged.

The patent position of biotechnology and pharmaceutical companies generally is highly uncertain, involves complex legal and factual questions and has in recent years been the subject of much litigation. The degree of patent protection we require to successfully commercialize our product candidates may be unavailable or severely limited in some cases and may not adequately protect our rights or permit us to gain or keep any competitive advantage. We cannot provide any assurances that any of our owned or in-licensed patents have, or that any of our owned or in-licensed pending patent applications that mature into issued patents will have, claims with a scope sufficient to protect rilonacept, mavrilimumab, KPL-716 or our other product candidates. In addition, the laws of foreign countries may not protect our rights to the same extent as the laws of the United States. Furthermore, patents have a limited lifespan. In the United States, the natural expiration of a patent is generally 20 years after it is filed. Various extensions and adjustments may be available; however, the life of a patent, and the protection it affords, is limited. The actual protection afforded by a patent varies on a product-by-product basis, from country-to-country and depends upon many factors, including the type of patent, the scope of its coverage, the availability of regulatory-related extensions, the availability of legal remedies in a particular country and the validity and enforceability of the patent.

Patents may be eligible for limited patent term extension in the United States under the Drug Price Competition and Patent Term Restoration Act of 1984, referred to as the Hatch-Waxman Act. Similar patent extensions exist in the European Union and Japan, subject to the applicable laws in those jurisdictions. We may not receive an extension if we fail to apply within applicable deadlines or fail to apply prior to expiration of relevant patents. For example, no patent term extension was obtained in the United States following the FDA's approval of rilonacept for the treatment of CAPS, and the deadline for applying for such extension has passed. Accordingly, patent term extension in the United States based on the FDA's approval of rilonacept for CAPS, or any other indication for which the FDA may grant approval in the future, is unavailable. Moreover, the length of the extension could be less than we request. If we are unable to obtain patent term extension or the term of any such extension is less than we request, the period during which we can enforce our patent rights for that product will be shortened and our competitors may obtain approval to market competing products sooner, impacting our revenue.

Given the amount of time required for the development, testing and regulatory review of new product candidates, patents protecting such candidates might expire before or shortly after such candidates are commercialized, thereby limiting protection such patent would afford the respective product and any competitive advantage such patent may provide. In some cases, an in-licensed patent portfolio may have undergone a considerable loss of patent term prior to our initiation of development and commercialization of the product candidate. For example, the patents covering rilonacept as a composition of matter have a term that expires in 2019 in the United States, not including patent term adjustment, and in 2023 in Europe, not including any patent term extensions. As a result, our owned and in-licensed patent portfolio may not provide us with adequate and continuing patent protection sufficient to exclude others from commercializing products similar or identical to our product candidates. In such cases, we expect to rely on regulatory exclusivity for our product candidates, such as orphan drug exclusivity, which generally grants seven years of marketing exclusivity under the Federal Food, Drug, and Cosmetic Act, and up to 10 years of marketing exclusivity in Europe. While, we expect to seek orphan drug designation for rilonacept in

the United States for the treatment of recurrent pericarditis, we may not be successful in obtaining such designation or we may not be able to maintain the benefits of the designation. Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent FDA from approving the same or a different drug in another indication. Even after an orphan drug is approved, the FDA can subsequently approve a later application for the same drug for the same condition if the FDA concludes that the later drug is clinically superior in that it is shown to be safer in a substantial portion of the target populations, more effective or makes a major contribution to patient care. In addition, a designated orphan drug may not receive orphan drug exclusivity if it is approved for a use that is broader than the indication for which it received orphan designation. See "— We may seek orphan drug designation for some of our product candidates and we may be unsuccessful, or may be unable to maintain the benefits associated with orphan drug designation, including the potential for market exclusivity, for any product candidate for which we obtain orphan drug designation."

Other parties may have developed or may develop technologies that may be related or competitive to our own, and such parties may have filed or may file patent applications, or may have received or may receive patents, claiming inventions that may overlap or conflict with those claimed in our patent applications or issued patents, with respect to either the same methods or formulations or the same subject matter, in either case, that we may rely upon to dominate our patent position in the market. Publications of discoveries in the scientific literature often lag behind the actual discoveries, and patent applications in the United States and other jurisdictions are typically not published until 18 months after filing, or in some cases not at all. Therefore, we cannot know with certainty whether we or our licensors were the first to make the inventions claimed in our owned or licensed patents or pending patent applications, or that we or our licensors were the first to file for patent protection of such inventions. As a result, the issuance, scope, validity, enforceability and commercial value of our patent rights cannot be predicted with any certainty.

In addition, the patent prosecution process is expensive and time-consuming, and we may not be able to file and prosecute all necessary or desirable patent applications at a reasonable cost or in a timely manner. Patent prosecution is a lengthy process, during which the scope of the claims initially submitted for examination by the USPTO is often significantly narrowed by the time they issue, if at all. The claims of our issued patents or patent applications when issued may not cover our product candidates, proposed commercial technologies or the future products that we develop, or even if such patents provide coverage, the coverage obtained may not provide any competitive advantage. It is also possible that we will fail to identify patentable aspects of our research and development output before it is too late to obtain patent protection. Moreover, in some circumstances, we do not have the right to control the preparation, filing and prosecution of patent applications, or to maintain the patents, covering technology that we license from third parties. Therefore, these patents and applications may not be prosecuted and enforced in a manner consistent with the best interests of our business. In the case of our field limited license from Regeneron, another licensee may have the right to enforce patents covering the product in their field. As a result, we may need to coordinate enforcement with another party, and the other party could enforce the patents in a manner adverse to our interests or otherwise put the patents at risk of invalidation.

The strength of patents in the biotechnology and pharmaceutical field involves complex legal and scientific questions and can be uncertain. The patent applications that we own or license may fail to result in issued patents in the United States or in foreign countries. Even if we acquire patent protection that we expect should enable us to maintain a competitive advantage, third parties may challenge the validity, enforceability or scope thereof, which may result in such patents being

narrowed, invalidated or held unenforceable. The issuance of a patent is not conclusive as to its inventorship, scope, validity, enforceability or term, and our owned and licensed patents may be challenged in the courts or patent offices in the United States and abroad. For example, we may be subject to a third-party submission of prior art to the USPTO challenging the priority of an invention claimed within one of our patents, which submissions may also be made prior to a patent's issuance, precluding the granting of any of our pending patent applications. We may become involved in contested proceedings challenging our patent rights or the patent rights of others from whom we have obtained licenses to such rights. For example, patents granted by the USPTO may be subject to third-party challenges such as (without limitation) derivation, re-examination, interference, post-grant review or *inter partes* review proceedings, and patents granted by the European Patent Office may be challenged by any person in an opposition proceeding within nine months from the publication of the grant. Similar proceedings are available in other jurisdictions, and in some jurisdictions third parties can raise questions of validity with a patent office even before a patent has granted. Competitors may claim that they invented the inventions claimed in our issued patents or patent applications prior to us, or may file patent applications before we do. In such case, we may have to participate in interference or derivation proceedings in the USPTO, to determine which party is entitled to a patent on the disputed invention. We may also become involved in similar opposition proceedings in the European Patent Office or similar offices in other jurisdictions regarding our intellectual property rights with respect to our products and technology.

Such proceedings can be expensive, time consuming and may divert the efforts of our technical and managerial personnel, which could in turn harm our business, whether or not we receive a determination favorable to us. We may not be able to correctly estimate or control our future operating expenses in relation to such proceedings, which could affect operating expenses. Our operating expenses may fluctuate significantly in the future as a result of a variety of factors, including the costs of such proceedings.

Since patent applications are confidential for a period of time after filing, we cannot be certain that we or our licensors were the first to file any patent application related to our product candidates. Competitors may also contest our patents, if issued, by showing the patent examiner that the invention was not original, was not novel or was obvious. In litigation, a competitor could claim that our patents, if issued, are not valid for a number of reasons. If a court agrees, rights to those challenged patents may be diminished or lost.

In addition, we may in the future be subject to claims by our former employees or consultants asserting an ownership right in our patents or patent applications, as a result of the work they performed on our behalf. Although we generally require all of our employees and consultants and any other partners or collaborators who have access to our proprietary know-how, information or technology to assign or grant similar rights to their inventions to us, we cannot be certain that we have executed such agreements with all parties who may have contributed to our intellectual property, nor can we be certain that our agreements with such parties will be upheld in the face of a potential challenge, or that they will not be breached, for which we may not have an adequate remedy.

An adverse determination in any such submission or proceeding may result in loss of exclusivity or freedom to operate or in patent claims being narrowed, invalidated or held unenforceable, in whole or in part, which could limit our ability to stop others from using or commercializing similar or identical technology and products, without payment to us, or could limit the duration of the patent protection covering our technology and product candidates. Such challenges may also result in our inability to manufacture or commercialize our product candidates without infringing third-party patent rights. In addition, if the breadth or strength of protection provided by our patents and patent applications is threatened, it could dissuade companies from collaborating with us to license, develop or commercialize current or future product candidates.

Even if they are unchallenged, our issued patents and our pending patent applications, if issued, may not provide us with any meaningful protection or prevent competitors from designing around our patent claims to circumvent our owned or licensed patents by developing similar or alternative technologies or products in a non-infringing manner. For example, a third-party may develop a competitive drug that provides benefits similar to one or more of our product candidates but that has a different composition that falls outside the scope of our patent protection. If the patent protection provided by the patents and patent applications we hold or pursue with respect to our product candidates is not sufficiently broad to impede such competition, or if the breadth, strength or term (including any extensions or adjustments) of protection provided by the patents and patent applications we hold or pursue with respect to our product candidates or any future product candidates is successfully challenged, our ability to successfully commercialize our product candidates could be negatively affected, which would harm our business. Further, if we encounter delays in our clinical trials, the period of time during which we could market our product candidates or any future product candidates under patent protection would be reduced.

Licensing of intellectual property is of critical importance to our business and involves complex legal, business and scientific issues. If we breach any of the agreements under which we acquired our product candidates, we could lose the ability to continue the development and commercialization of the related product. Additionally, our current licensing and acquisition agreements contain limitations and restrictions that could limit or adversely affect our ability to develop and commercialize other products in the future.

We entered into agreements to acquire the rights to develop and commercialize our product candidates, rilonacept, mavrilimumab, KPL-716, KPL-045 and KPL-404. In September 2017, we entered into a license agreement with Regeneron to obtain an exclusive license under certain intellectual property rights controlled by Regeneron to develop and commercialize rilonacept. In December 2017, we entered into a license agreement with MedImmune to obtain exclusive worldwide rights to research, develop, manufacture, market and sell mavrilimumab and any other products covered by the licensed patent rights. In September 2016, pursuant to an asset purchase agreement with Biogen, we acquired all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716, including patents and other intellectual property rights, clinical data, know-how and inventory. Each of these agreements requires us to use commercially reasonable efforts to develop and commercialize the related product candidates, make timely milestone and other payments, provide certain information regarding our activities with respect to such product candidates and indemnify the other party with respect to our development and commercialization activities under the terms of the agreements. In addition, we licensed KPL-045 from Novo Nordisk in August 2017 and the right to conduct research and development of KPL-404 from Primatope in September 2017. These current agreements and any future such agreements that we enter into impose a variety of obligations.

We are currently a party to a number of license and acquisition agreements of importance to our business and to our current product candidates, and we expect to be subject to additional such agreements in the future. Disputes may arise between us and any of these counterparties regarding intellectual property subject to and each parties' obligations under such agreements, including:

- our diligence obligations with respect to the use of the licensed technology in relation to our development and commercialization of our product candidates, and what activities satisfy those diligence obligations;
- the scope of rights granted under the agreement and other interpretation-related issues;
- our obligations to make milestone, royalty or other payments under those agreements;
- whether and the extent to which our technology and processes infringe on intellectual property of the licensor that is not subject to the agreement;

- our right to sublicense patent and other rights to third parties;
- the ownership of inventions and know-how resulting from the joint creation or use of intellectual property by our licensors and us and our partners;
- our right to transfer or assign the license; and
- the effects of termination.

These or other disputes over our obligations or intellectual property that we have licensed or acquired may prevent or impair our ability to maintain our current arrangements on acceptable terms, or may impair the value of the arrangement to us. Any such dispute could have an adverse affect on our business.

If we fail to meet our obligations under these agreements in a material respect, the respective licensor/seller would have the right to terminate the respective agreement and upon the effective date of such termination, have the right to re-obtain the related technology as well as aspects of any intellectual property controlled by us and developed during the period the agreement was in force that relate to the applicable technology. This means that the licensor/seller to each of these agreements could effectively take control of the development and commercialization of our product candidates after an uncured, material breach of the agreement by us. This would also be the case if we voluntarily terminate the relevant agreement. While we would expect to exercise all rights and remedies available to us, including seeking to cure any breach by us, and otherwise seek to preserve our rights under the technology licensed to or acquired by us, we may not be able to do so in a timely manner, at an acceptable cost or at all. Any uncured, material breach under the license could result in our loss of exclusive rights and may lead to a complete termination of our product development and any commercialization efforts for each of our product candidates.

Regeneron has rights to develop rilonacept in its retained fields of local administration to the eye and ear, oncology, deficiency of the interleukin-1 receptor, or DIRA, and CAPS. Regeneron may also develop rilonacept in fields to which we have licensed the rights, but we retain the commercial benefit related to that development upon approval of rilonacept in any field that we have licensed. We and Regeneron communicate with each other concerning our related development activities, and we have approval rights over Regeneron's development in the fields that we have licensed, including pericarditis. Outside of the United States and Japan, Regeneron has granted a third-party licensee the right to develop and commercialize rilonacept in CAPS and certain periodic fever syndromes. The development of rilonacept in other fields could increase the possibility of identification of adverse safety results that impact our development of rilonacept for recurrent pericarditis. In addition, if approved, commercialization of rilonacept in other fields could result in an increased threat of off-label use to compete with the sale of rilonacept to treat these indications, which may diminish sales of rilonacept in fields licensed exclusively to us.

Certain of our agreements may limit or delay our ability to consummate certain transactions, may impact the value of those transactions, or may limit our ability to pursue certain activities. For example, under the MedImmune Agreement, we cannot sublicense the rights licensed or sublicensed to us without the consent of MedImmune and certain applicable third-party licensors, if required by agreements between MedImmune and such third-party licensors. Under an asset purchase agreement with Biogen, or the Biogen Agreement, Biogen has a right of first negotiation under certain circumstances to purchase the assets we acquired from Biogen or to obtain a license to exploit the applicable products. This right of first negotiation remains in effect until the earlier of 12 years from the date of the agreement or the first commercial sale of a product under the agreement, and applies to a variety of transactions, including licensing transactions and the sale of our company. In addition, under the Biogen Agreement, we are subject to an exclusivity obligation, pursuant to which we may not conduct any activity alone or through a third party related to a

product that modulates OSMR (other than for the development and commercialization of products that are the subject of the Biogen Agreement). This exclusivity obligation runs from the earlier of the eighth anniversary of the agreement or the first commercial sale of a product that is the subject of the agreement.

Third parties may initiate legal proceedings alleging that we are infringing their intellectual property rights, the outcome of which would be uncertain and could have a material adverse effect on the success of our business.

Our commercial success depends upon our ability and the ability of our collaborators to develop, manufacture, market and sell our product candidates and use our proprietary technologies without infringing the proprietary rights and intellectual property of third parties. The biotechnology and pharmaceutical industries are characterized by extensive and frequent litigation regarding patents and other intellectual property rights. We cannot assure you that our product candidates or any future product candidates, including methods of making or using these product candidates, will not infringe existing or future third-party patents. We may in the future become party to, or threatened with, adversarial proceedings or litigation regarding intellectual property rights with respect to our product candidates and technology, including contested proceedings before the USPTO. Our competitors or other third parties may assert infringement claims against us, alleging that our products are covered by their patents.

Given the vast number of patents in our field of technology, we cannot be certain that we do not infringe existing patents or that we will not infringe patents that may be granted in the future. Many companies have filed, and continue to file, patent applications related to immunomodulation. Some of these patent applications have already been allowed or issued, and others may issue in the future. For example, we are aware of third-party patents that contain claims potentially relevant to certain therapeutic uses of mavrilimumab and KPL-716. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to mavrilimumab and KPL-716 would be found to infringe any valid claim of these patents. While we may decide to initiate proceedings to challenge the validity of these or other patents in the future, we may be unsuccessful, and courts or patent offices in the United States and abroad could uphold the validity of any such patent. If we were to challenge the validity of any issued United States patent in court, we would need to overcome a statutory presumption of validity that attaches to every United States patent. This means that in order to prevail, we would have to present clear and convincing evidence as to the invalidity of the patent's claims. In order to avoid infringing these or any other third-party patents, we may find it necessary or prudent to obtain licenses to such patents from such third-party intellectual property holders. However, we may be unable to secure such licenses or otherwise acquire or in-license any compositions, methods of use, processes, or other intellectual property rights from third parties that we identify as necessary for our current or future product candidates. The licensing or acquisition of third-party intellectual property rights is a competitive area, and several more established companies may also pursue strategies to license or acquire third-party intellectual property rights that we may consider attractive or necessary. These established companies may have a competitive advantage over us due to their size, capital resources and greater clinical development and commercialization capabilities. In addition, companies that perceive us to be a competitor may be unwilling to assign or license rights to us. We also may be unable to license or acquire third-party intellectual property rights on terms that would allow us to make an appropriate return on our investment or at all. If we are unable to successfully obtain rights to required third-party intellectual property rights or maintain the existing intellectual property rights we have, we may have to abandon development of the relevant program or product candidate, which could have a material adverse effect on our business, financial condition, results of operations and prospects.

Since our product candidates are being developed for use in fields that are competitive and of strong interest to pharmaceutical and biotechnology companies, we will likely seek to file additional patent applications and may have additional patents granted in the future, based on our future research and development efforts. Furthermore, because patent applications can take many years to issue and may be confidential for 18 months or more after filing, and because pending patent claims can be revised before issuance, there may be applications now pending which may later result in issued patents that may be infringed by the manufacture, use or sale of our product candidates. Regardless of when filed, we may fail to identify relevant third-party patents or patent applications, or we may incorrectly conclude that a third-party patent is invalid or not infringed by our product candidates or activities. If a patent holder believes our product candidate infringes its patent, the patent holder may sue us even if we have received patent protection for our technology. Moreover, we may face patent infringement claims from non-practicing entities that have no relevant drug revenue and against whom our own patent portfolio may thus have no deterrent effect. If a patent infringement suit were threatened or brought against us, we could be forced to stop or delay research, development, manufacturing or sales of the product candidate that is the subject of the actual or threatened suit.

If we are found to infringe a third-party's intellectual property rights, we could be required to obtain a license from such third-party to continue developing and marketing our product candidates and technology. Under any such license, we would most likely be required to pay various types of fees, milestones, royalties or other amounts. However, we may not be able to obtain any required license on commercially reasonable terms or at all. Even if we were able to obtain such a license, it could be granted on non-exclusive terms, thereby providing our competitors and other third parties access to the same technologies licensed to us. Without such a license, we could be forced, including by court order, to cease developing and commercializing the infringing technology or product candidate, or forced to redesign it, or to cease some aspect of our business operations. In addition, we could be found liable for monetary damages, including treble damages and attorneys' fees if we are found to have willfully infringed such third-party patent rights. We may be required to indemnify collaborators or contractors against such claims. A finding of infringement could prevent us from commercializing our product candidates or force us to cease some of our business operations, which could materially harm our business. Even if we are successful in defending against such claims, litigation can be expensive and time consuming and would divert management's attention from our core business. Any of these events could harm our business significantly.

We may become involved in lawsuits to protect or enforce our patents and other intellectual property rights, which could be expensive, time consuming and unsuccessful.

Competitors and other third parties may infringe, misappropriate or otherwise violate our patents and other intellectual property rights, whether owned or in-licensed. To counter infringement or unauthorized use, we or our current or future collaborators may be required to file infringement claims against these infringers. A court may disagree with our allegations, however, and may refuse to stop the other party from using the technology at issue on the grounds that our patents do not cover the third-party technology in question. Further, such third parties could counterclaim that we infringe their intellectual property or that a patent we have asserted against them is invalid or unenforceable. In patent litigation in the United States, defendant counterclaims challenging the validity, enforceability or scope of asserted patents are commonplace. In addition, third parties may initiate legal proceedings against us to assert such challenges to our intellectual property rights. The outcome of any such proceeding is generally unpredictable. Grounds for a validity challenge could be an alleged failure to meet any of several statutory requirements, including lack of novelty, obviousness or non-enablement. Patents may be unenforceable if someone connected with prosecution of the patent withheld relevant information from the USPTO or made a misleading

statement during prosecution. It is possible that prior art of which we or our licensors and the patent examiner were unaware during prosecution exists, which could render our patents invalid. Moreover, it is also possible that prior art may exist that we are aware of but do not believe is relevant to our current or future patents, but that could nevertheless be determined to render our patents invalid.

Some of our competitors may be able to devote significantly more resources to intellectual property litigation, and may have significantly broader patent portfolios to assert against us if we assert our rights against them. Further, because of the substantial discovery required in connection with intellectual property litigation, there is a risk that some of our confidential information could be disclosed or otherwise compromised during litigation.

An adverse result in any litigation proceeding could put one or more of our patents, whether owned or in-licensed, at risk of being invalidated or interpreted narrowly. If a defendant were to prevail on a legal assertion of invalidity or unenforceability of our patents covering one of our product candidates, we would lose at least part, and perhaps all, of the patent protection covering such product candidate. Competing products may also be sold in other countries in which our patent coverage might not exist or be as strong. If we lose a foreign patent lawsuit, alleging our infringement of a competitor's patents, we could be prevented from marketing our products in one or more foreign countries. Any of these outcomes would have a materially adverse effect on our business.

Intellectual property litigation could cause us to spend substantial resources and distract our personnel from their normal responsibilities.

Litigation or other legal proceedings relating to intellectual property claims, with or without merit, is unpredictable and generally expensive and time consuming and is likely to divert significant resources from our core business, including distracting our technical and management personnel from their normal responsibilities. Furthermore, because of the substantial amount of discovery required in connection with intellectual property litigation, there is a risk that some of our confidential information could be compromised by disclosure during this type of litigation. In addition, there could be public announcements of the results of hearings, motions or other interim proceedings or developments and if securities analysts or investors perceive these results to be negative, it could have a substantial adverse effect on the price of our Class A common shares. Such litigation or proceedings could substantially increase our operating losses and reduce the resources available for development activities or any future sales, marketing or distribution activities.

We may not have sufficient financial or other resources to adequately conduct such litigation or proceedings. Some of our competitors may be able to sustain the costs of such litigation or proceedings more effectively than we can because of their greater financial resources and more mature and developed intellectual property portfolios. Accordingly, despite our efforts, we may not be able to prevent third parties from infringing upon or misappropriating or from successfully challenging our intellectual property rights. Uncertainties resulting from the initiation and continuation of patent litigation or other proceedings could have a material adverse effect on our ability to compete in the marketplace.

Obtaining and maintaining patent protection depends on compliance with various procedural, document submission, fee payment and other requirements imposed by governmental patent agencies, and our patent protection could be reduced or eliminated for non-compliance with these requirements.

The USPTO and various foreign governmental patent agencies require compliance with a number of procedural, documentary, fee payment and other similar provisions during the patent

application process. In addition, periodic maintenance fees on issued patents often must be paid to the USPTO and foreign patent agencies over the lifetime of the patent. While an unintentional lapse can in many cases be cured by payment of a late fee or by other means in accordance with the applicable rules, there are situations in which noncompliance can result in abandonment or lapse of the patent or patent application, resulting in partial or complete loss of patent rights in the relevant jurisdiction. Non-compliance events that could result in abandonment or lapse of a patent or patent application include, but are not limited to, failure to respond to official actions within prescribed time limits, non-payment of fees and failure to properly legalize and submit formal documents. If we or our licensors fail to maintain the patents and patent applications covering our products or technologies, we may not be able to stop a competitor from marketing products that are the same as or similar to our product candidates, which would have a material adverse effect on our business. In addition, if we fail to apply for applicable patent term extensions or adjustments, we will have a more limited time during which we can enforce our granted patents. In addition, if we are responsible for patent prosecution and maintenance of patent rights in-licensed to us, any of the foregoing could expose us to liability to the applicable patent owner.

We may not be able to effectively enforce our intellectual property rights throughout the world.

Filing, prosecuting and defending patents on our product candidates in all countries throughout the world would be prohibitively expensive. The requirements for patentability may differ in certain countries, particularly in developing countries. Moreover, our ability to protect and enforce our intellectual property rights may be adversely affected by unforeseen changes in foreign intellectual property laws. In addition, the patent laws of some foreign countries do not afford intellectual property protection to the same extent as the laws of the United States. Many companies have encountered significant problems in protecting and defending intellectual property rights in certain foreign jurisdictions. Varying filing dates in international countries may also permit intervening third parties to allege priority to patent applications claiming certain technology. The legal systems of some countries, particularly developing countries, do not favor the enforcement of patents and other intellectual property rights. This could make it difficult for us to stop the infringement of our patents or the misappropriation of our other intellectual property rights. For example, many foreign countries have compulsory licensing laws under which a patent owner must grant licenses to third parties. In addition, many countries limit the enforceability of patents against certain parties, including government agencies or government contractors. Consequently, we may not be able to prevent third parties from practicing our inventions in all countries outside the United States. Competitors may use our technologies in jurisdictions where we have not obtained patent protection to develop their own products and, further, may export otherwise infringing products to territories where we have patent protection, if our ability to enforce our patents to stop infringing activities is inadequate. These products may compete with our product candidates, and our patents or other intellectual property rights may not be effective or sufficient to prevent them from competing.

Proceedings to enforce our patent rights, whether owned or in-licensed, in foreign jurisdictions, whether or not successful, could result in substantial costs and divert our efforts and resources from other aspects of our business. Furthermore, while we intend to pursue protection for our intellectual property rights in the major markets for our product candidates, we cannot ensure that we will be able to initiate or maintain similar efforts in all jurisdictions in which we may wish to market our product candidates. Accordingly, our efforts to protect our intellectual property rights in such countries may be inadequate. In addition, changes in the law and legal decisions by courts in the United States and foreign countries may affect our ability to obtain and enforce adequate intellectual property protection for our technology.

Changes to the patent law in the United States and other jurisdictions could diminish the value of patents in general, thereby impairing our ability to protect our product candidates.

As is the case with other biopharmaceutical companies, our success is heavily dependent on intellectual property, particularly patents. Obtaining and enforcing patents in the biopharmaceutical industry involve both technological and legal complexity and is therefore costly, time consuming and inherently uncertain. Patent reform legislation in the United States and other countries, including the Leahy-Smith America Invents Act, or Leahy-Smith Act, signed into law on September 16, 2011, could increase those uncertainties and costs. The Leahy-Smith Act includes a number of significant changes to U.S. patent law. These include provisions that affect the way patent applications are prosecuted, redefine prior art and provide more efficient and cost-effective avenues for competitors to challenge the validity of patents. In addition, the Leahy-Smith Act has transformed the U.S. patent system into a first-to-file system. The first-to-file provisions, however, only became effective on March 16, 2013. Accordingly, it is not yet clear what, if any, impact the Leahy-Smith Act will have on the operation of our business. However, the Leahy-Smith Act and its implementation could make it more difficult to obtain patent protection for our inventions, whether owned or in-licensed, and increase the uncertainties and costs surrounding the prosecution of our patent applications and the enforcement or defense of our issued patents, in each case whether owned or in-licensed, all of which could harm our business, results of operations and financial condition.

Among some of the other changes introduced by the Leahy-Smith Act are changes that limit where a patentee may file a patent infringement suit and provide new opportunities for third parties to challenge issued patents in the USPTO. We may be subject to the risk of third-party prior art submissions on pending applications or become a party to opposition, derivation, reexamination, *inter partes* review, post-grant review or interference proceedings challenging our patents. There is a lower standard of evidence necessary to invalidate a patent claim in a USPTO proceeding relative to the standard in U.S. district or federal court. This could lead third parties to challenge and successfully invalidate our patents that would not otherwise be invalidated if challenged through the court system.

The U.S. Supreme Court has ruled on several patent cases in recent years, either narrowing the scope of patent protection available in certain circumstances or weakening the rights of patent owners in certain situations. In addition, there have been recent proposals for additional changes to the patent laws of the United States and other countries that, if adopted, could impact our ability to obtain or maintain patent protection for our proprietary technology or our ability to enforce our proprietary technology. Depending on future actions by the U.S. Congress, the U.S. courts, the USPTO and the relevant law-making bodies in other countries, the laws and regulations governing patents could change in unpredictable ways that would weaken our ability to obtain new patents; enforce or shorten the term of our existing patents and patents that we might obtain in the future; shorten the term that has been lengthened by patent term adjustment of our existing patents or patents that we might obtain in the future; or challenge the validity or enforceability of patents that may be asserted against us by our competitors or other third parties.

If we are unable to protect the confidentiality of our trade secrets, our business and competitive position may be harmed.

In addition to the protection afforded by patents, we may rely upon unpatented trade secret protection, unpatented know-how and continuing technological innovation to develop and maintain our competitive position. Although we seek to protect our proprietary technology and processes, in part, by entering into confidentiality agreements with our collaborators, scientific advisors, contractors, employees and consultants, and invention assignment agreements with our consultants, scientific advisors and employees, we may not be able to prevent the unauthorized

disclosure or use of our technical know-how or other trade secrets by the parties to these agreements. Moreover, we cannot guarantee that we have entered into such agreements with each party that may have or have had access to our confidential information or proprietary technology and processes. Monitoring unauthorized uses and disclosures is difficult, and we do not know whether the steps we have taken to protect our proprietary technologies will be effective. If any of the collaborators, scientific advisors, employees, contractors and consultants who are parties to these agreements breaches or violates the terms of any of these agreements, we may not have adequate remedies for any such breach or violation, and we could lose our trade secrets as a result. Moreover, if confidential information that is licensed or disclosed to us by our partners, collaborators, or others is inadvertently disclosed or subject to a breach or violation, we may be exposed to liability to the owner of that confidential information. Enforcing a claim that a third-party illegally obtained and is using our trade secrets, like patent litigation, is expensive and time consuming, and the outcome is unpredictable. In addition, courts outside the United States are sometimes less willing to protect trade secrets.

We cannot be certain that the steps we have taken will prevent unauthorized use or unauthorized reverse engineering of our technology. Monitoring unauthorized use of our intellectual property is difficult and costly. We may not be able to detect unauthorized use of, or take appropriate steps to enforce, our intellectual property rights. The steps we have taken to protect our proprietary rights may not be adequate to prevent misappropriation of our intellectual property.

We also seek to preserve the integrity and confidentiality of our data and other confidential information by maintaining physical security of our premises and physical and electronic security of our information technology systems. While we have confidence in these individuals, organizations and systems, agreements or security measures may be breached and detecting the disclosure or misappropriation of confidential information and enforcing a claim that a party illegally disclosed or misappropriated confidential information is difficult, expensive and time-consuming, and the outcome is unpredictable. Further, we may not be able to obtain adequate remedies for any breach. In addition, our confidential information may otherwise become known or be independently discovered by competitors, in which case we would have no right to prevent them, or those to whom they communicate it, from using that technology or information to compete with us. We may in the future rely on trade secret protection, which would be subject to the risks identified above with respect to confidential information.

Our trade secrets could otherwise become known or be independently discovered by our competitors. Competitors could purchase our product candidates and attempt to replicate some or all of the competitive advantages we derive from our development efforts, willfully infringe our intellectual property rights, design around our protected technology or develop their own competitive technologies that fall outside of our intellectual property rights. If any of our trade secrets were to be lawfully obtained or independently developed by a competitor, we would have no right to prevent them, or those to whom they communicate it, from using that technology or information to compete with us. If our trade secrets are not adequately protected so as to protect our market against competitors' products, our competitive position could be adversely affected, as could our business.

If our trademarks and trade names are not adequately protected, then we may not be able to build name recognition in our markets of interest and our business may be adversely affected.

Our registered or unregistered trademarks or trade names may be challenged, infringed, circumvented or declared generic or determined to be infringing on other marks. We may not be able to protect our rights to these trademarks and trade names, which we need to build name recognition among potential partners or customers in our markets of interest. At times, competitors or other third parties may adopt trade names or trademarks similar to ours, thereby impeding our

ability to build brand identity and possibly leading to market confusion. In addition, there could be potential trade name or trademark infringement claims brought by owners of other registered trademarks or trademarks that incorporate variations of our registered or unregistered trademarks or trade names. Over the long term, if we are unable to establish name recognition based on our trademarks and trade names, then we may not be able to compete effectively and our business may be adversely affected. Our efforts to enforce or protect our proprietary rights related to trademarks, trade secrets, domain names, copyrights or other intellectual property may be ineffective and could result in substantial costs and diversion of resources and could adversely affect our business, financial condition, results of operations and prospects.

We have not yet registered trademarks for a commercial trade name for our lead product candidates in the United States or foreign jurisdictions and failure to secure such registrations could adversely affect our business.

We have not yet registered trademarks for a commercial trade name for some of our lead product candidates in the United States or any foreign jurisdiction. During trademark registration proceedings, we may receive rejections. Although we are given an opportunity to respond to those rejections, we may be unable to overcome such rejections. In addition, in the USPTO and in comparable agencies in many foreign jurisdictions, third parties are given an opportunity to oppose pending trademark applications and to seek to cancel registered trademarks. Opposition or cancellation proceedings may be filed against our trademarks, and our trademarks may not survive such proceedings. Moreover, any name we propose to use with our product candidates in the United States must be approved by the FDA, regardless of whether we have registered it, or applied to register it, as a trademark. The FDA typically conducts a review of proposed product names, including an evaluation of potential for confusion with other product names. If the FDA objects to any of our proposed proprietary product names, we may be required to expend significant additional resources in an effort to identify a suitable substitute name that would qualify under applicable trademark laws, not infringe the existing rights of third parties and be acceptable to the FDA.

Risks Related to Commercialization

The incidence and prevalence for target patient populations of our product candidates have not been established with precision. If the market opportunities for our product candidates are smaller than we estimate, or if any approval that we obtain is based on a narrower definition of the patient population, our revenue and ability to achieve profitability may be materially adversely affected.

The precise incidence and prevalence for all the conditions we aim to address with our programs are unknown. Our projections of both the number of people who have these diseases, as well as the subset of people with these diseases who have the potential to benefit from treatment with our product candidates, are based on beliefs and estimates. These estimates have been derived from a variety of sources, including the scientific literature, surveys of clinics, patient foundations or market research, and may prove to be incorrect. Further, new trials may change the estimated incidence or prevalence of these diseases. The number of patients may turn out to be lower than expected. We estimate that there are approximately:

- 3,000 to 12,000 addressable patients not well-managed with existing therapies with recurrent pericarditis in the United States;
- 100,000 to 200,000 prevalent patients in the United States with GCA, with similar rates in other major markets;

- 300,000 addressable patients with prurigo nodularis in the United States, of which we believe 20% to 30% to have severe systemic disease with similar prevalence rates in other major markets; and
- 300,000 addressable patients with moderate-to-severe atopic dermatitis in the United States with similar rates in other major markets.

The total addressable market across all of our product candidates will ultimately depend upon, among other things, the diagnosis criteria included in the final label for each of our product candidates approved for sale for these indications, acceptance by the medical community and patient access, drug pricing and reimbursement. The number of patients in the United States and other major markets and elsewhere may turn out to be lower than expected, patients may not be otherwise amenable to treatment with our products or new patients may become increasingly difficult to identify or gain access to, all of which would adversely affect our results of operations and our business. Further, even if we obtain significant market share for our product candidates, because the potential target populations are very small, we may never achieve profitability despite obtaining such significant market share.

If, in the future, we are unable to establish our own sales, marketing and distribution capabilities, or enter into agreements with third parties to sell and market our product candidates, we may not be successful in commercializing our product candidates if and when they are approved, and we may not be able to generate any revenue.

We do not currently have a sales, marketing or distribution infrastructure. We have never sold, marketed or distributed any therapeutic products. To achieve commercial success for any approved product candidate, we must build our sales, marketing, managerial and other non-technical capabilities or make arrangements with third parties to perform these services. We currently plan to establish our own sales and marketing capabilities and directly commercialize any approved product candidate.

There are risks involved with both establishing our own sales and marketing capabilities and entering into arrangements with third parties to perform these services. For example, recruiting and training a sales force is expensive and time consuming and could delay any drug launch. If the commercial launch of a product candidate for which we recruit a sales force and establish marketing capabilities is delayed or does not occur for any reason, we would have prematurely or unnecessarily incurred these commercialization expenses. This may be costly, and our investment would be lost if we cannot retain or reposition our sales and marketing personnel.

Factors that may inhibit our efforts to commercialize our product candidates on our own include:

- our inability to recruit and retain adequate numbers of effective sales and marketing personnel;
- the inability of sales personnel to obtain access to physicians or persuade adequate numbers of physicians to prescribe any future products;
- the lack of complementary products to be offered by sales personnel, which may put us at a competitive disadvantage relative to companies with more extensive product lines;
- our inability to equip medical and sales personnel with effective materials, including medical and sales literature to help them educate physicians and other healthcare providers regarding applicable diseases and our future products;
- our inability to develop or obtain sufficient operational functions to support our commercial activities; and

- unforeseen costs and expenses associated with creating an independent sales and marketing organization.

If we enter into arrangements with third parties to perform sales, marketing, distribution and other commercial support services, our product revenues or the profitability of these revenues to us are likely to be lower than if we were to market and sell any product candidates that we develop ourselves. In addition, we may not be successful in entering into arrangements with third parties to sell and market our product candidates or may be unable to do so on terms that are favorable to us. We likely will have little control over such third parties, and any of them may fail to devote the necessary resources and attention to sell and market our product candidates effectively. Developing a sales and marketing organization requires significant investment, is time consuming and could delay the launch of our product candidates. We may not be able to build an effective sales and marketing organization in the United States, the European Union or other key global markets. If we do not establish sales and marketing capabilities successfully, either on our own or in collaboration with third parties, we will not be successful in commercializing our product candidates. Further, our business, results of operations, financial condition and prospects will be materially adversely affected.

Our current or future product candidates may not gain market acceptance by physicians or patients, in which case our ability to generate product revenues will be compromised.

Even if the FDA or any other regulatory authority approves the marketing of our product candidates, whether developed on our own or with a collaborator, physicians, healthcare providers, patients or the medical community may not accept or use our product candidates. If our product candidates do not achieve an adequate level of acceptance, we may not generate significant product revenue or any profits from operations. The degree of market acceptance of our product candidates will depend on a variety of factors, including:

- the timing of market introduction;
- the number and clinical profile of competing products;
- the potential and perceived advantages of our product candidates over alternative treatments;
- the clinical indications for which our product candidates are approved;
- our ability to provide acceptable evidence of safety and efficacy;
- the prevalence and severity of any side effects;
- product labeling or product insert requirements of the FDA or other regulatory authorities;
- limitations or warnings contained in the labeling approved by the FDA or other regulatory authorities;
- relative convenience and ease of administration, including as compared to alternative treatments and competitive therapies;
- cost-effectiveness, particularly in relation to alternative treatments;
- the effectiveness of our sales, marketing and distribution support;
- availability of adequate coverage, reimbursement and payment from health maintenance organizations and other insurers, both public and private; and
- other potential advantages over alternative treatment methods.

If our product candidates fail to gain market acceptance, our ability to generate revenues will be adversely affected. Even if our product candidates achieve market acceptance, the market may prove not to be large enough to allow us to generate significant revenues.

The successful commercialization of our product candidates will depend in part on the extent to which governmental authorities and health insurers establish adequate coverage, reimbursement levels and pricing policies. Failure to obtain or maintain coverage and adequate reimbursement for our product candidates, if approved, could limit our ability to market those products and decrease our ability to generate revenue.

Our ability to commercialize any product candidates successfully also will depend in part on the extent to which adequate coverage and reimbursement for these product candidates and related treatments will be available from government authorities, private health insurers and other organizations. Government authorities and third-party payors, such as private health insurers and health maintenance organizations, decide which products they will pay for and establish reimbursement levels. A primary trend in the U.S. healthcare industry and elsewhere is cost containment. Government authorities and third-party payors have attempted to control costs by limiting coverage and the amount of reimbursement for particular products. Increasingly, third-party payors are requiring that drug companies provide them with predetermined discounts from list prices and are challenging the prices charged for products. We cannot be sure that adequate coverage will be available for any product candidate that we commercialize and, if coverage is available, that the level of reimbursement will be adequate or that will not require co-payments that patients may find unacceptably high. Reimbursement may impact the demand for, or the price of, any product candidate for which we obtain marketing approval. If reimbursement is not available or is available only to limited levels, we may not be able to successfully commercialize any product candidate for which we obtain marketing approval. Any coverage or reimbursement that may become available may be decreased or eliminated in the future.

There may be significant delays in obtaining reimbursement for newly approved products, and coverage may be more limited than the purposes for which the drug is approved by the FDA or similar regulatory authorities outside the United States. Moreover, eligibility for reimbursement does not imply that any drug will be paid for in all cases or at a rate that covers our costs, including research, development, manufacture, sale and distribution. Interim reimbursement levels for new products, if applicable, may also not be sufficient to cover our costs and may not be made permanent. Reimbursement rates may vary according to the use of the product and the clinical setting in which it is used, may be based on reimbursement levels already set for lower-cost products and may be incorporated into existing payments for other services. Net prices for products may be reduced by mandatory discounts or rebates required by government healthcare programs or private payors and by any future relaxation of laws that presently restrict imports of products from countries where they may be sold at lower prices than in the United States. Third-party payors often rely upon Medicare coverage policy and payment limitations in setting their own reimbursement policies. Our inability to promptly obtain coverage and profitable payment rates from both government-funded and private payors for any approved drugs that we develop could have a material adverse effect on our operating results, our ability to raise capital needed to commercialize drugs and our overall financial condition.

There is significant uncertainty related to the insurance coverage and reimbursement of newly-approved products. In the United States, third-party payors, including private and governmental payors, such as the Medicare and Medicaid programs, play an important role in determining the extent to which new drugs and biologics will be covered. The Medicare and Medicaid programs increasingly are used as models in the United States for how private payors and other governmental payors develop their coverage and reimbursement policies for drugs and biologics.

Some third-party payors may require pre-approval of coverage for new or innovative drug therapies before they will reimburse healthcare providers who use such therapies. It is difficult to predict at this time what third-party payors will decide with respect to the coverage and reimbursement for our product candidates.

Third-party payors increasingly are challenging prices charged for pharmaceutical or biologic products and services, and many third-party payors may refuse to provide coverage and reimbursement for particular drugs or biologics when an equivalent generic drug, biosimilar or a less expensive therapy is available. It is possible that a third-party payor may consider our product candidates as substitutable and only offer to reimburse patients for the less expensive product. Even if we show improved efficacy or improved convenience of administration with our product candidates, pricing of existing products may limit the amount we will be able to charge for our product candidates. These payors may deny or revoke the reimbursement status of a given product or establish prices for new or existing marketed products at levels that are too low to enable us to realize an appropriate return on our investment in our product candidates. If reimbursement is not available or is available only at limited levels, we may not be able to successfully commercialize our product candidates, and may not be able to obtain a satisfactory financial return on our product candidates.

The regulations that govern regulatory approvals, pricing and reimbursement for new products vary widely from country to country. Our operations are subject to extensive governmental price control or other market regulations in other countries outside of the United States, and we believe the increasing emphasis on cost-containment initiatives in European and other countries have and will continue to put pressure on the pricing and usage of our product candidates. Some countries require approval of the sale price of a drug before it can be marketed. In many countries, the pricing review period begins after marketing approval is granted. In some foreign markets, prescription pharmaceutical pricing remains subject to continuing governmental control even after initial approval is granted. As a result, we might obtain marketing approval for a product candidate in a particular country, but then be subject to price regulations that delay our commercial launch of the product candidate, possibly for lengthy time periods, and negatively impact the revenues we are able to generate from the sale of the product candidate in that country. Adverse pricing limitations may hinder our ability to recoup our investment in one or more product candidates, even if our product candidates obtain marketing approval.

Moreover, increasing efforts by governmental and third-party payors in the United States and abroad to cap or reduce healthcare costs may cause such organizations to limit both coverage and the level of reimbursement for newly approved products and, as a result, they may not cover or provide adequate payment for our product candidates. We expect to experience pricing pressures in connection with the sale of our product candidates due to the trend toward managed healthcare, the increasing influence of health maintenance organizations and additional legislative changes. The downward pressure on healthcare costs in general, particularly prescription drugs and biologics and surgical procedures and other treatments, has become intense. As a result, increasingly high barriers are being erected to the entry of new products.

Our future growth may depend, in part, on our ability to penetrate foreign markets, where we would be subject to additional regulatory burdens and other risks and uncertainties.

Our future profitability may depend, in part, on our ability to commercialize our product candidates in foreign markets for which we may rely on collaboration with third parties. Although we do not have immediate plans to pursue the commercialization of rilonacept for recurrent pericarditis outside of the United States, we are evaluating the opportunities for the development and commercialization of our product candidates in foreign markets. We are not permitted to market or promote any of our product candidates before we receive regulatory approval from the applicable

regulatory authority in that foreign market, and we may never receive such regulatory approval for any of our product candidates. To obtain separate regulatory approval in many other countries we must comply with numerous and varying regulatory requirements of such countries regarding safety and efficacy and governing, among other things, clinical trials and commercial sales, pricing and distribution of our product candidates, and we cannot predict success in these jurisdictions. If we obtain approval of our product candidates and ultimately commercialize our product candidates in foreign markets, we would be subject to additional risks and uncertainties, including:

- our customers' ability to obtain reimbursement for our product candidates in foreign markets;
- our inability to directly control commercial activities because we are relying on third parties;
- the burden of complying with complex and changing foreign regulatory, tax, accounting and legal requirements;
- different medical practices and customs in foreign countries affecting acceptance in the marketplace;
- import or export licensing requirements;
- longer accounts receivable collection times;
- longer lead times for shipping;
- language barriers for technical training and the need for language translations;
- reduced protection of intellectual property rights in some foreign countries;
- the existence of additional potentially relevant third-party intellectual property rights;
- foreign currency exchange rate fluctuations; and
- the interpretation of contractual provisions governed by foreign laws in the event of a contract dispute.

Foreign sales of our product candidates could also be adversely affected by the imposition of governmental controls, political and economic instability, trade restrictions and changes in tariffs.

In some countries, particularly the countries in Europe, the pricing of prescription pharmaceuticals is subject to governmental control. In these countries, pricing negotiations with governmental authorities can take considerable time after the receipt of marketing approval for a drug. To obtain reimbursement or pricing approval in some countries, we may be required to conduct a clinical trial that compares the cost-effectiveness of our product candidate to other available therapies. If reimbursement of our products is unavailable or limited in scope or amount, or if pricing is set at unsatisfactory levels, our business could be harmed, possibly materially.

Even if we receive regulatory approval for any of our product candidates, we will be subject to ongoing obligations and continued regulatory review, which may result in significant additional expense. Additionally, our product candidates, if approved, could be subject to labeling and other restrictions and market withdrawal and we may be subject to penalties if we fail to comply with regulatory requirements or experience unanticipated problems with our products.

If the FDA or a comparable foreign regulatory authority approves any of our product candidates, it or they will be subject to ongoing regulatory requirements for manufacturing, labeling, packaging, distribution, storage, advertising, promotion, sampling, record-keeping adverse event reporting, conduct of post-marketing trials and submission of safety, efficacy and other post-market information, including both federal and state requirements in the United States and requirements of comparable foreign regulatory authorities.

Manufacturers and manufacturers' facilities are required to comply with extensive FDA, and comparable foreign regulatory authority, requirements, including ensuring that quality control and manufacturing procedures conform to cGMP regulations. As such, we and our CMOs will be subject to user fees and continual review and inspections to assess compliance with cGMP and adherence to commitments made in any BLA or MAA. Accordingly, we and others with whom we work must continue to expend time, money and effort in all areas of regulatory compliance, including manufacturing, production and quality control.

Any regulatory approvals that we receive for our product candidates may be subject to limitations on the approved indicated uses for which the product may be marketed or to the conditions of approval, or contain requirements for potentially costly post-marketing testing, including Phase 4 clinical trials, and surveillance to monitor the safety and efficacy of the product candidate. We will be required to report certain adverse reactions and production problems, if any, to the FDA and comparable foreign regulatory authorities. Any new legislation addressing drug safety issues could result in delays in product development or commercialization, or increased costs to assure compliance.

We will have to comply with requirements concerning advertising and promotion for our products. Promotional communications with respect to prescription drugs and biologics are subject to a variety of legal and regulatory restrictions and must be consistent with the information in the product's approved label. As such, we may not promote our products for indications or uses for which they do not have approval. The holder of an approved BLA or MAA must submit new or supplemental applications and obtain approval for certain changes to the approved product, product labeling or manufacturing process. We could also be asked to conduct post-marketing clinical trials to verify the safety and efficacy of our products in general or in specific patient subsets. If original marketing approval were obtained via the accelerated approval pathway, we could be required to conduct a successful post-marketing clinical trial to confirm clinical benefit for our products. An unsuccessful post-marketing trial or failure to complete such a trial could result in the withdrawal of marketing approval. The FDA also may place other conditions on approvals including the requirement for a REMS, to assure the safe use of the product. If the FDA concludes a REMS is needed, the sponsor of the BLA must submit a proposed REMS before it can obtain approval. A REMS could include medication guides, physician communication plans or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools.

If a regulatory agency discovers previously unknown problems with a product, such as adverse events of unanticipated severity or frequency, or problems with the facility where the product is manufactured, or disagrees with the promotion, marketing or labeling of a product, such regulatory agency may impose restrictions on that product or us, including requiring withdrawal of the product from the market. If we discover previously unknown problems with a product candidate, including adverse events of unanticipated severity or frequency, or with our manufacturing processes, or fail to comply with regulatory requirements, a regulatory agency or enforcement authority may, among other things:

- issue warning letters;
- impose civil or criminal penalties;
- suspend or withdraw regulatory approval;
- suspend any of our ongoing clinical trials;
- refuse to approve pending applications or supplements to approved applications submitted by us;

- impose restrictions on our operations, including closing our CMOs' facilities; or
- seize or detain products, or require a product recall.

Any government investigation of alleged violations of law could require us to expend significant time and resources in response, and could generate negative publicity. Any failure to comply with ongoing regulatory requirements may significantly and adversely affect our ability to commercialize and generate revenue from our products. If regulatory sanctions are applied or if regulatory approval is withdrawn, the value of our company and our operating results will be adversely affected.

If there are changes in the application of legislation or regulatory policies, or if problems are discovered with a product or the manufacture of a product, or if we or one of our distributors, licensees or co-marketers fails to comply with regulatory requirements, the regulatory authorities could take various actions. These include imposing fines on us, imposing restrictions on our product or its manufacture and requiring us to recall or remove a product from the market. The regulatory authorities could also suspend or withdraw our marketing authorizations, or require us to conduct additional clinical trials, change our product labeling or submit additional applications for marketing authorization. If any of these events occurs, our ability to sell our product may be impaired, and we may incur substantial additional expense to comply with regulatory requirements.

The policies of the FDA and other regulatory authorities may change and additional government regulations may be enacted that could prevent, limit or delay regulatory approval of our product candidates. We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative or executive action, either in the United States, Europe or in other jurisdictions. For example, the current U.S. presidential administration has taken several executive actions, including the issuance of a number of Executive Orders, that could impose significant burdens on, or otherwise materially delay, the FDA's ability to engage in routine oversight activities such as implementing statutes through rulemaking, issuance of guidance and review and approval of marketing applications. It is difficult to predict how these Executive Orders will be implemented, and the extent to which they will impact the FDA's ability to exercise its regulatory authority. If these executive actions impose restrictions on the FDA's ability to engage in oversight and implementation activities in the normal course, our business may be negatively impacted. In addition, if we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we are not able to maintain regulatory compliance, we may lose any marketing approval that we may have obtained and we may not achieve or sustain profitability.

Our business operations and current and future relationships with investigators, healthcare professionals, consultants, customers and third-party payors will be subject to applicable anti-kickback, fraud and abuse, physician payment transparency, health information privacy and security and other healthcare laws and regulations, which could expose us to criminal sanctions, civil penalties, exclusion from government healthcare programs, contractual damages, reputational harm and diminished profits and future earnings.

Although we do not currently have any products on the market, once we begin commercializing our product candidates, if approved, we will be subject to additional healthcare statutory and regulatory requirements and enforcement by the federal government and the states and foreign governments in which we conduct our business. Healthcare providers, physicians and third-party payors will play a primary role in the recommendation and prescription of any product candidates for which we obtain marketing approval. Our future arrangements with third-party payors and customers may expose us to broadly applicable fraud and abuse and other healthcare laws and regulations that may constrain the business or financial arrangements and relationships

through which we market, sell and distribute our product candidates for which we obtain marketing approval. Restrictions under applicable federal and state healthcare laws and regulations, include the following:

- the U.S. federal Anti-Kickback Statute, which prohibits, among other things, persons from knowingly and willfully soliciting, offering, receiving or providing remuneration, directly or indirectly, in cash or in kind, to induce or reward either the referral of an individual for, or the purchase, order or recommendation of, any good or service, for which payment may be made under federal and state healthcare programs such as Medicare and Medicaid. A person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation. The U.S. federal Anti-Kickback Statute has been interpreted to apply to arrangements between pharmaceutical manufacturers on the one hand and prescribers, purchasers and formulary managers on the other hand;

- the U.S. federal False Claims Act and civil monetary penalties laws, which, among other things, impose criminal and civil penalties, including through civil whistleblower or qui tam actions, against individuals or entities for knowingly presenting, or causing to be presented, to the federal government, claims for payment that are false or fraudulent or making a false statement to avoid, decrease or conceal an obligation to pay money to the federal government. In addition, the government may assert that a claim including items and services resulting from a violation of the federal Anti-Kickback Statute constitutes a false of fraudulent claim for purposes of the False Claims Act. As a result of a modification made by the Fraud Enforcement and Recovery Act of 2009, a claim includes "any request or demand" for money or property presented to the federal government. In addition, manufacturers can be held liable under the FCA even when they do not submit claims directly to government payors if they are deemed to "cause" the submission of false or fraudulent claims;

- the U.S. federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, which imposes criminal and civil liability for executing a scheme to defraud any healthcare benefit program or obtain, by means of false or fraudulent pretenses, representations or promises, any of the money or property owned by, or under the custody or control of, any healthcare benefit program, regardless of the payor (e.g., public or private) and knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false statement in connection with the delivery of or payment for healthcare benefits, items or services; similar to the federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation;

- the U.S. federal physician payment transparency requirements, sometimes referred to as the "Sunshine Act" created under Section 60002 of the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act, or the Affordable Care Act, which requires manufacturers of drugs, devices, biologics and medical supplies that are reimbursable under Medicare, Medicaid or the Children's Health Insurance Program to report to the Department of Health and Human Services information related to physician payments and other transfers of value and the ownership and investment interests of such physicians and their immediate family members;

- HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act of 2009, or HITECH, and its implementing regulations, which also imposes obligations on certain covered entity healthcare providers, health plans and healthcare clearinghouses as well as their business associates that perform certain services involving the use or disclosure of individually identifiable health information, including mandatory contractual

terms, with respect to safeguarding the privacy, security and transmission of individually identifiable health information; and

- analogous state laws and regulations, such as state anti-kickback and false claims laws that may apply to sales or marketing arrangements and claims involving healthcare items or services reimbursed by non-governmental third-party payors, including private insurers; and some state laws require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government in addition to requiring drug manufacturers to report information related to payments to physicians and other healthcare providers or marketing expenditures, and state laws governing the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and often are not preempted by HIPAA, thus complicating compliance efforts.

These laws and regulations, among other things, may constrain our business, marketing and other promotional activities by limiting the kinds of financial arrangements we may have with hospitals, physicians or other potential purchasers of our product candidates, if approved. We have entered into consulting and advisory board agreements with physicians, some of whom are paid in the form of shares or options to acquire our common shares. We could be adversely affected if regulatory agencies determine our financial relationships with such physicians to be in violation of applicable laws. Because of the breadth of these laws and the narrowness of the statutory exceptions and regulatory safe harbors available under such laws, it is possible that some of our business activities could be subject to challenge under one or more of such laws. The scope and enforcement of each of these laws is uncertain and subject to rapid change in the current environment of healthcare reform, especially in light of the lack of applicable precedent and regulations.

Interactions between biopharmaceutical companies and physicians are also governed by strict laws, regulations, industry self-regulation codes of conduct and physicians' codes of professional conduct in the individual EU member states. The provision of any inducements to physicians to prescribe, recommend, endorse, order, purchase, supply, use or administer a drug product is prohibited. A number of EU member states have established additional rules requiring pharmaceutical companies to publicly disclose their interactions with physicians and to obtain approval from employers, professional organizations and/or competent authorities before entering into agreements with physicians.

Ensuring that our future business arrangements with third parties comply with applicable healthcare laws and regulations will involve substantial costs. It is possible that governmental authorities will conclude that our business practices do not comply with current or future statutes, regulations or case law involving applicable fraud and abuse or other healthcare laws and regulations. If our operations, including anticipated activities to be conducted by our sales team, were to be found to be in violation of any of these laws or any other governmental regulations that may apply to us, we may be subject to the imposition of civil, criminal and administrative penalties, damages, disgorgement, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, individual imprisonment, contractual damages, reputational harm, diminished profits and future earnings, additional reporting requirements and/or oversight if we become subject to a corporate integrity agreement or similar agreement to resolve allegations of non-compliance with these laws, and curtailment or restructuring of our operations, any of which could adversely affect our ability to operate our business and our results of operations. Further, defending against any such actions can be costly, time-consuming and may require significant personnel resources. Therefore, even if we are successful in defending against any such actions that may be brought against us, our business may be impaired.

Product liability lawsuits against us could cause us to incur substantial liabilities and could limit commercialization of any product candidates that we may develop.

We will face an inherent risk of product liability exposure related to the testing of our product candidates in human clinical trials and will face an even greater risk if we commercially sell any product candidates that we may develop. If we cannot successfully defend ourselves against claims that our product candidates caused injuries, we could incur substantial liabilities. Regardless of merit or eventual outcome, liability claims may result in:

- decreased demand for any product candidates that we may develop;
- injury to our reputation and significant negative media attention;
- regulatory investigations that could require costly recalls or product modifications;
- withdrawal of clinical trial participants;
- significant costs to defend the related litigation;
- substantial monetary awards to trial participants or patients;
- loss of potential revenue;
- the diversion of management's attention away from managing our business; and
- the inability to commercialize any product candidates that we may develop.

Although we maintain product liability insurance coverage, it may not be adequate to cover all liabilities that we may incur and is subject to deductibles and coverage limitations. We anticipate that we will need to increase our insurance coverage when and if we successfully commercialize any product candidate. Insurance coverage is increasingly expensive. We may not be able to maintain insurance coverage at a reasonable cost or in an amount adequate to satisfy any liability that may arise. If we are unable to obtain insurance at acceptable cost or otherwise protect against potential product liability claims, we will be exposed to significant liabilities, which may materially and adversely affect our business and financial position. These liabilities could prevent or interfere with our commercialization efforts.

Other Risks Related to Our Business

Enacted and future healthcare legislation may have a material adverse effect on our business and results of operations.

In the United States, European Union and other jurisdictions, there have been and we expect there will continue to be a number of legislative and regulatory initiatives and proposed changes to the healthcare system that could affect our future operations. For example, in the United States, in March 2010, the Affordable Care Act was passed, which substantially changes the way healthcare is financed by both governmental and private insurers, and significantly impacts the U.S. pharmaceutical industry. The Affordable Care Act, among other things, subjects biologic products to potential competition by lower-cost biosimilars, addresses a new methodology by which rebates owed by manufacturers under the Medicaid Drug Rebate Program are calculated for drugs and biologics that are inhaled, infused, instilled, implanted or injected, increases the minimum Medicaid rebates owed by manufacturers under the Medicaid Drug Rebate Program and extends the rebate program to individuals enrolled in Medicaid managed care organizations, establishes annual fees and taxes on manufacturers of certain branded prescription drugs and biologics, including our product candidates, and a new Medicare Part D coverage gap discount program, in which manufacturers must agree to offer 50% point-of-sale discounts which, through subsequent legislative amendments, was increased to 70%, off negotiated prices of applicable brand drugs and

biologics to eligible beneficiaries during their coverage gap period, as a condition for the manufacturer's outpatient products to be covered under Medicare Part D.

Since its enactment, there have been judicial and Congressional challenges to certain aspects of the Affordable Care Act. The current Presidential Administration and U.S. Congress have attempted and will likely continue to seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Most recently, the Tax Cuts and Jobs Act was enacted, which, among other things, removes penalties for not complying with the Affordable Care Act's individual mandate to carry health insurance. It is uncertain the extent to which any such changes may impact our business or financial condition.

In addition, other legislative changes have been proposed and adopted in the United States since the Affordable Care Act was enacted. On August 2, 2011, the Budget Control Act of 2011, among other things, led to aggregate reductions of Medicare payments to providers of 2% per fiscal year. These reductions went into effect on April 1, 2013 and, due to subsequent legislative amendments to the statute, will remain in effect through 2025 unless additional Congressional action is taken. On January 2, 2013, the American Taxpayer Relief Act of 2012 was signed into law, which, among other things, further reduced Medicare payments to several types of providers.

Additionally, there has been increasing legislative and enforcement interest in the United States with respect to specialty drug pricing practices. Specifically, there have been several recent U.S. Congressional inquiries and proposed federal and state legislation designed to, among other things, bring more transparency to drug and biologic pricing, reduce the cost of prescription drugs and biologics under Medicare, review the relationship between pricing and manufacturer patient programs and reform government program reimbursement methodologies for drugs and biologics. Moreover, payment methodologies may be subject to changes in healthcare legislation and regulatory initiatives. We expect that additional U.S. federal healthcare reform measures will be adopted in the future, any of which could limit the amounts that the U.S. federal government will pay for healthcare products and services, which could result in reduced demand for our product candidates or additional pricing pressures.

Individual states in the United States have also become increasingly active in passing legislation and implementing regulations designed to control pharmaceutical and biological product pricing, including price or patient reimbursement constraints, discounts, restrictions on certain product access and marketing cost disclosure and transparency measures, and, in some cases, designed to encourage importation from other countries and bulk purchasing. Legally-mandated price controls on payment amounts by third-party payors or other restrictions could harm our business, results of operations, financial condition and prospects. In addition, regional healthcare authorities and individual hospitals are increasingly using bidding procedures to determine what pharmaceutical products and which suppliers will be included in their prescription drug and other healthcare programs. This could reduce the ultimate demand for our product candidates or put pressure on our product pricing.

In the European Union, similar political, economic and regulatory developments may affect our ability to profitably commercialize our product candidates, if approved. In addition to continuing pressure on prices and cost containment measures, legislative developments at the EU or member state level may result in significant additional requirements or obstacles that may increase our operating costs. The delivery of healthcare in the European Union, including the establishment and operation of health services and the pricing and reimbursement of medicines, is almost exclusively a matter for national, rather than EU, law and policy. National governments and health service providers have different priorities and approaches to the delivery of healthcare and the pricing and reimbursement of products in that context. In general, however, the healthcare budgetary constraints in most EU member states have resulted in restrictions on the pricing and

reimbursement of medicines by relevant health service providers. Coupled with ever-increasing EU and national regulatory burdens on those wishing to develop and market products, this could prevent or delay marketing approval of our product candidates, restrict or regulate post-approval activities and affect our ability to commercialize our product candidates, if approved.

In markets outside of the United States and European Union, reimbursement and healthcare payment systems vary significantly by country, and many countries have instituted price ceilings on specific products and therapies.

We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative action, either in the United States, the European Union or elsewhere. If we or any third-party we may engage are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we or such third-party are not able to maintain regulatory compliance, our product candidates may lose any regulatory approval that may have been obtained and we may not achieve or sustain profitability.

Unfavorable global economic conditions could adversely affect our business, financial condition or results of operations.

Our results of operations could be adversely affected by general conditions in the global economy and in the global financial markets. For example, the global financial crisis caused extreme volatility and disruptions in the capital and credit markets. A severe or prolonged economic downturn, such as the global financial crisis, could result in a variety of risks to our business, including, weakened demand for our product candidates and our ability to raise additional capital when needed on acceptable terms, if at all. A weak or declining economy could also strain our suppliers, possibly resulting in supply disruption, or cause our customers to delay making payments for our services. Doing business internationally involves a number of risks, including but not limited to:

- multiple, conflicting and changing laws and regulations such as privacy regulations, tax laws, export and import restrictions;
- employment laws, regulatory requirements and other governmental approvals, permits and licenses;
- failure by us to obtain and maintain regulatory approvals for the use of our products in various countries;
- additional potentially relevant third-party patent rights;
- complexities and difficulties in obtaining protection and enforcing our intellectual property;
- difficulties in staffing and managing foreign operations;
- complexities associated with managing multiple payor reimbursement regimes, government payors or patient self-pay systems;
- limits in our ability to penetrate international markets;
- financial risks, such as longer payment cycles, difficulty collecting accounts receivable, the impact of local and regional financial crises on demand and payment for our products and exposure to foreign currency exchange rate fluctuations;
- natural disasters, political and economic instability, including wars, terrorism, political unrest, outbreak of disease and boycotts;
- curtailment of trade, and other business restrictions;

- certain expenses including, among others, expenses for travel, translation and insurance; and
- regulatory and compliance risks that relate to maintaining accurate information and control over sales and activities that may fall within the purview of the U.S. Foreign Corrupt Practices Act, its books and records provisions or its anti-bribery provisions.

Any of the foregoing could harm our business and we cannot anticipate all of the ways in which the current economic climate and financial market conditions could adversely impact our business.

Our internal computer systems, or those of our third-party CMOs, CROs or other contractors or consultants, may fail or suffer security breaches, which could result in a material disruption of our product candidates' development programs.

Despite the implementation of security measures, our internal computer systems and those of our third-party CMOs, CROs and other contractors and consultants are vulnerable to damage from computer viruses, unauthorized access, theft, natural disasters, terrorism, war and telecommunication and electrical failures. While we have not experienced any such system failure, accident or security breach to date, if such an event were to occur and cause interruptions in our operations, it could result in a material disruption of our programs. For example, the loss of clinical trial data for our product candidates could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any disruption or security breach results in a loss of or damage to our data or applications or other data or applications relating to our technology or product candidates, or inappropriate disclosure or theft of confidential or proprietary information, we could incur liabilities and the further development of our product candidates could be delayed.

We face potential liability related to the privacy of health information we obtain from clinical trials sponsored by us.

Most healthcare providers, including research institutions from which we obtain patient health information, are subject to privacy and security regulations promulgated under HIPAA, as amended by the HITECH. We are not currently classified as a covered entity or business associate under HIPAA and thus is not subject to its requirements or penalties. However, any person may be prosecuted under HIPAAs criminal provisions either directly or under aiding-and-abetting or conspiracy principles. Consequently, depending on the facts and circumstances, we could face substantial criminal penalties if we knowingly receive individually identifiable health information from a HIPAA-covered healthcare provider or research institution that has not satisfied HIPAAs requirements for disclosure of individually identifiable health information. In addition, we may maintain sensitive personally identifiable information, including health information, that we receive throughout the clinical trial process, in the course of our research collaborations, and directly from individuals (or their healthcare providers) who enroll in our patient assistance programs. As such, we may be subject to state laws requiring notification of affected individuals and state regulators in the event of a breach of personal information, which is a broader class of information than the health information protected by HIPAA. Our clinical trial programs outside the United States may implicate international data protection laws, including the EU Data Protection Directive and legislation of the EU member states implementing it.

Our activities outside the United States impose additional compliance requirements and generate additional risks of enforcement for noncompliance. Failure by our CROs and other third-party contractors to comply with the strict rules on the transfer of personal data outside of the European Union into the United States may result in the imposition of criminal and administrative

sanctions on such collaborators, which could adversely affect our business. Furthermore, certain health privacy laws, data breach notification laws, consumer protection laws and genetic testing laws may apply directly to our operations and/or those of our collaborators and may impose restrictions on our collection, use and dissemination of individuals' health information. Moreover, patients about whom we or our collaborators obtain health information, as well as the providers who share this information with us, may have statutory or contractual rights that limit our ability to use and disclose the information. We may be required to expend significant capital and other resources to ensure ongoing compliance with applicable privacy and data security laws. Claims that we have violated individuals' privacy rights or breached our contractual obligations, even if we are not found liable, could be expensive and time-consuming to defend and could result in adverse publicity that could harm our business.

If we or third-party CMOs, CROs or other contractors or consultants fail to comply with applicable federal, state or local regulatory requirements, we could be subject to a range of regulatory actions that could affect our or our contractors' ability to develop and commercialize our product candidates and could harm or prevent sales of any affected products that we are able to commercialize, or could substantially increase the costs and expenses of developing, commercializing and marketing our products. Any threatened or actual government enforcement action could also generate adverse publicity and require that we devote substantial resources that could otherwise be used in other aspects of our business. Increasing use of social media could give rise to liability, breaches of data security or reputational damage.

We and our employees are increasingly utilizing social media tools as a means of communication both internally and externally.

Despite our efforts to monitor evolving social media communication guidelines and comply with applicable rules, there is risk that the use of social media by us or our employees to communicate about our product candidates or business may cause us to be found in violation of applicable requirements. In addition, our employees may knowingly or inadvertently make use of social media in ways that may not comply with our social media policy or other legal or contractual requirements, which may give rise to liability, lead to the loss of trade secrets or other intellectual property or result in public exposure of personal information of our employees, clinical trial patients, customers and others. Furthermore, negative posts or comments about us or our product candidates in social media could seriously damage our reputation, brand image and goodwill. Any of these events could have a material adverse effect on our business, prospects, operating results and financial condition and could adversely affect the price of our Class A common shares.

Our employees, principal investigators, CROs, consultants and other third-party service providers may engage in misconduct or other improper activities, including non-compliance with regulatory standards and requirements and insider trading.

We are exposed to the risk that our employees, principal investigators, CROs, consultants and other third-party service providers may engage in fraudulent conduct or other illegal activity. Misconduct by these parties could include intentional, reckless and/or negligent conduct or disclosure of unauthorized activities to us that violate the regulations of the FDA and other regulatory authorities, including those laws requiring the reporting of true, complete and accurate information to such authorities; healthcare fraud and abuse laws and regulations in the United States and abroad; or laws that require the reporting of financial information or data accurately.

In particular, sales, marketing and business arrangements in the healthcare industry are subject to extensive laws and regulations intended to prevent fraud, misconduct, kickbacks, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, including off-label promotion, sales

commission, customer incentive programs and other business arrangements. Activities subject to these laws also involve the improper use of information obtained in the course of clinical trials or creating fraudulent data in our pre-clinical studies or clinical trials, which could result in regulatory sanctions and cause serious harm to our reputation.

We intend to adopt, prior to the completion of this offering, a code of conduct applicable to all of our employees, but it is not always possible to identify and deter misconduct by employees and other third parties, and the precautions we take to detect and prevent this activity may not be effective in controlling unknown or unmanaged risks or losses or in protecting us from governmental investigations or other actions or lawsuits stemming from a failure to comply with these laws or regulations. Additionally, we are subject to the risk that a person could allege such fraud or other misconduct, even if none occurred. If any such actions are instituted against us, and we are not successful in defending ourselves or asserting our rights, those actions could have a significant impact on our business, including the imposition of civil, criminal and administrative penalties, damages, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, contractual damages, reputational harm, diminished profits and future earnings and curtailment of our operations, any of which could adversely affect our ability to operate our business and our results of operations.

We may acquire businesses, or products or product candidates, or form strategic alliances, in the future, and we may not realize the benefits of such acquisitions.

We have acquired and in-licensed, and may acquire or in-license additional businesses or products, from other companies or create joint ventures with third parties that we believe will complement or augment our existing business. If we acquire businesses with promising markets or technologies, we may not be able to realize the benefit of acquiring such businesses if we are unable to successfully integrate them with our existing operations and company culture. We may encounter numerous difficulties in developing, manufacturing and marketing any new products resulting from a strategic alliance or acquisition that delay or prevent us from realizing their expected benefits or enhancing our business. We cannot assure you that, following any such acquisition or license, we will achieve the expected synergies to justify the transaction.

Risks Related to Our Common Shares and This Offering

After this offering, our executive officers and certain other members of our senior management will have the ability to control all matters submitted to shareholders for approval.

Our Class A1 common shares and Class B1 common shares have no voting rights. As a result, all matters submitted to our shareholders will be decided by the vote of holders of our Class A common shares and Class B common shares. Each Class A common share is entitled to one vote per Class A common share and each Class B common share is entitled to ten votes per Class B common share. Following this offering, our executive officers and certain other members of our senior management will hold 72.6% of our voting power and have the ability to control the outcome of all matters submitted to our shareholders for approval. This concentrated control limits other shareholders' ability to influence corporate matters and may have an adverse effect on the price of our Class A common shares. As a result of the Class A common shares and Class B common shares that they will hold upon the closing of this offering, our executive officers and certain other members of our senior management will be able to control our management and affairs and the outcome of matters submitted to our shareholders for approval, including the election of directors and any sale, merger, consolidation or sale of all or substantially all of our assets. Our executive officers and certain other members of our senior management may have interests, with respect to their investment, that are different from our other investors, including the

investors in this offering. In addition, this concentration of ownership might adversely affect the market price of our Class A common shares by:

- delaying, deferring or preventing a change of control of us;
- impeding a merger, consolidation, takeover or other business combination involving us; or
- discouraging a potential acquirer from making a tender offer or otherwise attempting to obtain control of us.

In addition, each holder of Class B1 common shares has the ability to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time, and each holder of our Class A1 common shares has the ability to convert any portion of its Class A1 common shares into Class A common shares at any time. However, our Class A1 common shares and Class B1 common shares cannot be converted if, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of the issued and outstanding Class A common shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act outstanding unless such holders provide us with 61-days' prior notice that they intend to increase, decrease or waive such threshold upon conversion. Due to these conversion rights, holders of our Class A1 common shares and our Class B1 common shares could, at any time, significantly increase their voting control of us, which could result in their ability to significantly influence or control matters submitted to our shareholders for approval. For example, upon 61-days' prior written notice, entities affiliated with one of our directors, Felix J. Baker, could convert their Class A1 common shares and Class B1 common shares into Class A common shares and Class B common shares respectively which in the aggregate would result in such entities holding 73.8% of the voting power of our outstanding share capital following this offering.

The price of our Class A common shares is likely to be volatile and fluctuate substantially, which could result in substantial losses for purchasers of our Class A common shares in this offering.

Our share price is likely to be volatile. The shares market in general and the market for biopharmaceutical companies in particular have experienced extreme volatility that has often been unrelated to the operating performance of particular companies. As a result of this volatility, you may not be able to sell your Class A common shares at or above the initial public offering price. The market price for our Class A common shares may be influenced by many factors, including:

- the results of clinical trials for our product candidates;
- delays in in-licensing or acquiring additional complementary product candidates;
- delays in the commencement, enrollment and the ultimate completion of clinical trials;
- the results and potential impact of competitive products or technologies;
- our ability to manufacture and successfully produce our product candidates;
- actual or anticipated changes in estimates as to financial results, development timelines or recommendations by securities analysts;
- the level of expenses related to any of our product candidates or clinical development programs;
- variations in our financial results or those of companies that are perceived to be similar to us;
- financing or other corporate transactions, or inability to obtain additional funding;
- failure to meet or exceed expectations of the investment community;
- regulatory or legal developments in the United States and other countries;
- the recruitment or departure of key personnel;

- developments or disputes concerning patent applications, issued patents or other proprietary rights;
- the results of our efforts to discover, develop, acquire or in-license additional product candidates;
- changes in the structure of healthcare payment systems;
- market conditions in the pharmaceutical and biotechnology sectors;
- general economic, industry and market conditions;
- changes in voting control of our executive officers and certain other members of our senior management or affiliates who hold our shares; and
- the other factors described in this "Risk Factors" section.

An active trading market for our Class A common shares may not develop, and you may not be able to resell your shares at or above the initial public offering price.

Prior to this offering, there has been no public market for our Class A common shares. Although we anticipate that our Class A common shares will be approved for listing on The Nasdaq Global Market, an active trading market for our Class A common shares may never develop or be sustained following this offering. The initial public offering price of our Class A common shares will be determined through negotiations between us and the underwriters. This initial public offering price may not be indicative of the market price of our Class A common shares after this offering. In the absence of an active trading market for our Class A common shares, investors may not be able to sell their Class A common shares at or above the initial public offering price or at the time that they would like to sell.

If securities or industry analysts do not publish research or reports, or publish unfavorable research or reports, about us, our business or our market, our shares price and trading volume could decline.

The trading market for our Class A common shares will be influenced by the research and reports that equity research analysts publish about us and our business. We do not currently have and may never obtain research coverage by equity research analysts. Equity research analysts may elect not to provide research coverage of our Class A common shares after this offering, and such lack of research coverage may adversely affect the market price of our Class A common shares. In the event we do have equity research analyst coverage, we will not have any control over the analysts or the content and opinions included in their reports. The price of our shares could decline if one or more equity research analysts downgrades our shares or issues other unfavorable commentary or research. If one or more equity research analysts ceases coverage of our company or fails to publish reports on us regularly, demand for our Class A common shares could decrease, which in turn could cause the price of our Class A common shares or its trading volume to decline.

Sales of a substantial number of our Class A common shares in the public market could cause our share price to fall.

If our existing shareholders sell, or indicate an intention to sell, substantial amounts of our Class A common shares in the public market after the lock-up and other legal restrictions on resale discussed in this prospectus lapse, the market price of our Class A common shares could decline. Based upon the number of common shares outstanding as of March 31, 2018, upon the completion of this offering, we will have outstanding a total of 13,270,569 Class A common shares, 12,995,954 Class A1 common shares, 4,638,855 Class B common shares and 16,057,618 Class B1 common

shares assuming the conversion of all of our preferred shares into common shares upon the closing of this offering, no exercise of options to purchase Class A common shares outstanding as of March 31, 2018 and no exercise of the underwriters' option to purchase additional Class A common shares. Of these shares, only the Class A common shares sold in this offering, plus any Class A common shares sold upon exercise of the underwriters' option to purchase additional Class A common shares, will be freely tradable, without restriction, in the public market immediately following this offering.

Substantially all of our shareholders have entered into lock-up agreements pertaining to this offering with the underwriters that restrict their ability to sell or transfer their common shares, including common shares upon the conversion of preferred shares. The lock-up agreements will expire 180 days from the date of this prospectus. After the lock-up agreements expire, up to an additional 39,837,144 Class A common shares will be eligible for sale in the public market (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares). Approximately 27,551,086 of these Class A common shares will be held by our directors, executive officers and certain entities affiliated with our directors, and will, following the expiration of the lock-up, remain subject to certain limitations on sales made by affiliates pursuant to Rule 144 under the Securities Act. In addition, our Class A1 common shares, Class B common shares and Class B1 common shares automatically convert into Class A common shares upon transfer to non-affiliates. As a result, up to 28,449,865 of our Class A common shares may be issued upon such transfers. The representatives of the underwriters may, in their sole discretion, permit our officers, directors and other shareholders who are subject to these lock-up agreements to sell shares prior to the expiration of the lock-up.

Upon completion of this offering, 4,662,727 of our Class A common shares that are subject to outstanding options or reserved for future issuance under our equity incentive plans will become eligible for sale in the public market to the extent permitted by the provisions of various vesting schedules, the lock-up agreements and Rule 144 and Rule 701 under the Securities Act. If these additional Class A common shares are sold, or if it is perceived that they will be sold, in the public market, the market price of our Class A common shares could decline.

After this offering, the holders of approximately 35,670,093 of our Class A common shares (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares), will be entitled to rights with respect to the registration of their shares under the Securities Act, subject to the lock-up agreements described above. Registration of these shares under the Securities Act would result in the shares becoming freely tradable without restriction under the Securities Act, except for shares purchased by affiliates. Any sales of securities by these shareholders could have a material adverse effect on the market price of our Class A common shares.

If you purchase Class A common shares in this offering, you will suffer immediate dilution of your investment.

The initial public offering price of our Class A common shares is substantially higher than the as adjusted net tangible book value per common share. Therefore, if you purchase Class A common shares in this offering, you will pay a price per Class A common share that substantially exceeds our as adjusted net tangible book value per common share after this offering. To the extent outstanding options are exercised, you will incur further dilution. Based on the assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, you will experience immediate dilution of $11.21 per common share, representing the difference between our as adjusted net tangible book value per common share after giving effect to this offering and the assumed initial public offering price. In addition, purchasers of Class A common shares in this offering will have contributed approximately 28.2% of

the aggregate price paid by all purchasers of our common shares but will own only approximately 14.9% of our common shares outstanding after this offering. See "Dilution."

Future sales and issuances of our common shares or rights to purchase common shares, including pursuant to our equity incentive plans, could result in additional dilution of the percentage ownership of our shareholders and could cause our Class A common share price to fall.

We will need additional capital in the future to continue our planned operations. To the extent we raise additional capital by issuing equity securities, our shareholders may experience substantial dilution. We may sell common shares, convertible securities or other equity securities in one or more transactions at prices and in a manner we determine from time to time. If we sell common shares, convertible securities or other equity securities in more than one transaction, investors may be materially diluted by subsequent sales. These sales may also result in material dilution to our existing shareholders, and new investors could gain rights superior to our existing shareholders.

We have broad discretion in how we use the proceeds of this offering and may not use these proceeds effectively, which could affect our results of operations and cause our shares price to decline.

We will have considerable discretion in the application of the net proceeds of this offering. We intend to use the net proceeds from this offering, together with our existing cash and cash equivalents, to fund our clinical and pre-clinical development programs, working capital and other general corporate purposes. We may also use a portion of the net proceeds from this offering to in-license, acquire or invest in additional businesses, technologies, products or assets, although currently we have no specific agreements, commitments or understandings in this regard. As a result, investors will be relying upon management's judgment with only limited information about our specific intentions for the use of the balance of the net proceeds of this offering. We may use the net proceeds for purposes that do not yield a significant return or any return at all for our shareholders. In addition, pending their use, we may invest the net proceeds from this offering in a manner that does not produce income or that loses value.

We are an "emerging growth company," and the reduced disclosure requirements applicable to emerging growth companies may make our Class A common shares less attractive to investors.

We are an "emerging growth company," as defined in the JOBS Act. We will remain an emerging growth company until the earlier of (a) the last day of the fiscal year in which we have total annual gross revenues of $1.07 billion or more; (b) the last day of the fiscal year following the fifth anniversary of the date of the completion of this offering; (c) the date on which we have issued more than $1 billion in nonconvertible debt during the previous three years; or (d) the date on which we are deemed to be a large accelerated filer under the rules of the Securities and Exchange Commission, which means the market value of our Class A common shares that is held by non-affiliates exceeds $700 million as of the prior June 30th. For so long as we remain an emerging growth company, we are permitted and intend to rely on exemptions from certain disclosure requirements that are applicable to other public companies that are not emerging growth companies. These exemptions include:

- not being required to comply with the auditor attestation requirements of Section 404 of the Sarbanes-Oxley Act of 2002, or Section 404;

- an exemption from compliance with the requirement of the Public Company Accounting Oversight Board regarding the communication of critical audit matters in the auditor's report on the financial statements;
- providing only two years of audited financial statements in addition to any required unaudited interim financial statements and a correspondingly reduced "Management's Discussion and Analysis of Financial Condition and Results of Operations" disclosure;
- reduced disclosure obligations regarding executive compensation; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachute payments not previously approved. In this prospectus, we have not included all of the executive compensation-related information that would be required if we were not an emerging growth company.

We may choose to take advantage of some, but not all, of the available exemptions. We have taken advantage of reduced reporting burdens in this prospectus. In particular, we have provided only two years of audited financial statements and have not included all of the executive compensation information that would be required if we were not an emerging growth company. We cannot predict whether investors will find our Class A common shares less attractive if we rely on these exemptions. If some investors find our Class A common shares less attractive as a result, there may be a less active trading market for our Class A common shares and our shares price may be more volatile.

In addition, the JOBS Act provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. This allows an emerging growth company to delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of this exemption from new or revised accounting standards and, therefore, we will be subject to the same new or revised accounting standards as other public companies that are not emerging growth companies.

We will incur increased costs as a result of operating as a public company, and our management will be required to devote substantial time to new compliance initiatives.

As a public company, and particularly after we are no longer an "emerging growth company," we will incur significant legal, accounting and other expenses that we did not incur as a private company. In addition, the Sarbanes-Oxley Act of 2002 and rules subsequently implemented by the Securities and Exchange Commission and Nasdaq have imposed various requirements on public companies, including establishment and maintenance of effective disclosure and financial controls and corporate governance practices. Our management and other personnel will need to devote a substantial amount of time to these compliance initiatives. Moreover, these rules and regulations will increase our legal and financial compliance costs and will make some activities more time-consuming and costly. For example, we expect that these rules and regulations may make it more difficult and more expensive for us to obtain director and officer liability insurance.

Pursuant to Section 404, we will be required to furnish a report by our management on our internal control over financial reporting, including an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. However, while we remain an emerging growth company, we will not be required to include an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. To achieve compliance with Section 404 within the prescribed period, we will be engaged in a process to document and evaluate our internal control over financial reporting, which is both costly and challenging. In this regard, we will need to continue to dedicate internal resources, potentially

engage outside consultants and adopt a detailed work plan to assess and document the adequacy of internal control over financial reporting, continue steps to improve control processes as appropriate, validate through testing that controls are functioning as documented and implement a continuous reporting and improvement process for internal control over financial reporting. Despite our efforts, there is a risk that neither we nor our independent registered public accounting firm will be able to conclude within the prescribed timeframe that our internal control over financial reporting is effective as required by Section 404. This could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statements.

We have anti-takeover provisions in our amended and restated bye-laws that may discourage a change of control.

Our amended and restated bye-laws will contain provisions that could make it more difficult for a third-party to acquire us without the consent of our board of directors. These provisions will provide for:

- a classified board of directors with staggered three-year terms;
- directors only to be removed for cause;
- an affirmative vote of $66^{2}/_{3}\%$ of the voting power of our voting shares for certain "business combination" transactions that have not been approved by our board of directors;
- restrictions on the time period in which directors may be nominated; and
- our board of directors to determine the powers, preferences and rights of our preferred shares and to issue the preferred shares without shareholder approval.

These anti-takeover defenses could discourage, delay or prevent a transaction involving a change in control of our company and may prevent our shareholders from receiving the benefit from any premium to the market price of our Class A common shares offered by a bidder in a takeover context. Even in the absence of a takeover attempt, the existence of these provisions may adversely affect the prevailing market price of our Class A common shares if the provisions are viewed as discouraging takeover attempts in the future. These provisions could also discourage proxy contests, make it more difficult for you and other shareholders to elect directors of your choosing and cause us to take corporate actions other than those you desire. See "Description of Share Capital."

Because we do not anticipate paying any cash dividends on our capital shares in the foreseeable future, capital appreciation, if any, will be your sole source of gain.

We have never declared or paid cash dividends on our capital shares. We currently intend to retain all of our future earnings, if any, to finance the growth and development of our business. Additionally, the proposal to pay future dividends to shareholders will in addition effectively be at the sole discretion of our board of directors after taking into account various factors our board of directors deems relevant, including our business prospects, capital requirements, financial performance and new product development. As a result, capital appreciation, if any, of our Class A common shares will be your sole source of gain for the foreseeable future.

Risks Related to Owning Shares in a Bermuda Exempted Company and Certain Tax Risks

We are a Bermuda company and it may be difficult for you to enforce judgments against us or our directors and executive officers.

We are a Bermuda exempted company. As a result, the rights of holders of our Class A common shares will be governed by Bermuda law and our memorandum of association and

bye-laws. The rights of shareholders under Bermuda law may differ from the rights of shareholders of companies incorporated in other jurisdictions. It may be difficult for investors to enforce in the United States judgments obtained in U.S. courts against us based on the civil liability provisions of the U.S. securities laws. It is doubtful whether courts in Bermuda will enforce judgments obtained in other jurisdictions, including the United States, against us or our directors or officers under the securities laws of those jurisdictions or entertain actions in Bermuda against us or our directors or officers under the securities laws of other jurisdictions. See "Enforcement of Civil Liabilities Under United States Federal Securities Laws" for additional information.

Bermuda law differs from the laws in effect in the United States and may afford less protection to our shareholders.

We are organized under the laws of Bermuda. As a result, our corporate affairs are governed by the Bermuda Companies Act 1981, as amended, or the Companies Act, which differs in some material respects from laws typically applicable to U.S. corporations and shareholders, including the provisions relating to interested directors, amalgamations, mergers and acquisitions, takeovers, shareholder lawsuits and indemnification of directors. Generally, the duties of directors and officers of a Bermuda company are owed to the company only. Shareholders of Bermuda companies typically do not have rights to take action against directors or officers of the company and may only do so in limited circumstances. Shareholder class actions are not available under Bermuda law. The circumstances in which shareholder derivative actions may be available under Bermuda law are substantially more proscribed and less clear than they would be to shareholders of U.S. corporations. The Bermuda courts, however, would ordinarily be expected to permit a shareholder to commence an action in the name of a company to remedy a wrong to the company where the act complained of is alleged to be beyond the corporate power of the company or illegal, or would result in the violation of the company's memorandum of association or bye-laws. Furthermore, consideration would be given by a Bermuda court to acts that are alleged to constitute a fraud against the minority shareholders or, for instance, where an act requires the approval of a greater percentage of the company's shareholders than those who actually approved it.

When the affairs of a company are being conducted in a manner that is oppressive or prejudicial to the interests of some shareholders, one or more shareholders may apply to the Supreme Court of Bermuda, which may make such order as it sees fit, including an order regulating the conduct of the company's affairs in the future or ordering the purchase of the shares of any shareholders by other shareholders or by the company. Additionally, under our bye-laws and as permitted by Bermuda law, each shareholder has waived any claim or right of action against our directors or officers for any action taken by directors or officers in the performance of their duties, except for actions involving fraud or dishonesty. In addition, the rights of our shareholders and the fiduciary responsibilities of our directors under Bermuda law are not as clearly established as under statutes or judicial precedent in existence in jurisdictions in the United States, particularly the State of Delaware. Therefore, our shareholders may have more difficulty protecting their interests than would shareholders of a corporation incorporated in a jurisdiction within the United States.

There are regulatory limitations on the ownership and transfer of our common shares.

Common shares may be offered or sold in Bermuda only in compliance with the provisions of the Companies Act and the Bermuda Investment Business Act 2003, as amended, which regulates the sale of securities in Bermuda. In addition, the Bermuda Monetary Authority must approve all issues and transfers of shares of a Bermuda exempted company. However, the Bermuda Monetary Authority has, pursuant to its statement of June 1, 2005, given its general permission under the Exchange Control Act 1972 and related regulations for the issue and free transfer of our common shares to and among persons who are non-residents of Bermuda for exchange control purposes as

long as the shares are listed on an appointed shares exchange, which includes The Nasdaq Global Market. This general permission would cease to apply if we were to cease to be listed on The Nasdaq Global Market.

We may become subject to unanticipated tax liabilities.

Although we are incorporated under the laws of Bermuda, we may become subject to income, withholding or other taxes in certain jurisdictions by reason of our activities and operations, and it is also possible that taxing authorities in any such jurisdictions could assert that we are subject to greater taxation than we currently anticipate. Any such non-Bermudan tax liability could materially adversely affect our results of operations.

Taxing authorities could reallocate our taxable income among our subsidiaries, which could increase our overall tax liability.

We are incorporated under the laws of Bermuda and currently have a subsidiary in the United States. If we succeed in growing our business, we expect to conduct increased operations through our subsidiaries in various tax jurisdictions pursuant to transfer pricing arrangements between us, our parent company and our subsidiaries. If two or more affiliated companies are located in different countries, the tax laws or regulations of each country generally will require that transfer prices be the same as those between unrelated companies dealing at arms' length and that appropriate documentation is maintained to support the transfer prices. While we believe that we operate in compliance with applicable transfer pricing laws and intend to continue to do so, our transfer pricing procedures are not binding on applicable tax authorities.

If tax authorities in any of these countries were to successfully challenge our transfer prices as not reflecting arms' length transactions, they could require us to adjust our transfer prices and thereby reallocate our income to reflect these revised transfer prices, which could result in a higher tax liability to us. In addition, if the country from which the income is reallocated does not agree with the reallocation, both countries could tax the same income, resulting in double taxation. If tax authorities were to allocate income to a higher tax jurisdiction, subject our income to double taxation or assess interest and penalties, it would increase our consolidated tax liability, which could adversely affect our financial condition, results of operations and cash flows.

Changes in our effective tax rate may reduce our net income in future periods.

Our tax position could be adversely impacted by changes in tax rates, tax laws, tax practice, tax treaties or tax regulations or changes in the interpretation thereof by the tax authorities in Europe (including the United Kingdom), the United States, Bermuda and other jurisdictions, as well as being affected by certain changes currently proposed by the Organization for Economic Co-operation and Development and their action plan on Base Erosion and Profit Shifting. Such changes may become more likely as a result of recent economic trends in the jurisdictions in which we operate, particularly if such trends continue. If such a situation was to arise, it could adversely impact our tax position and our effective tax rate. Failure to manage the risks associated with such changes, or misinterpretation of the laws providing such changes, could result in costly audits, interest, penalties and reputational damage, which could adversely affect our business, results of our operations and our financial condition.

Our actual effective tax rate may vary from our expectation and that variance may be material. A number of factors may increase our future effective tax rates, including:

- the jurisdictions in which profits are determined to be earned and taxed;
- the resolution of issues arising from any future tax audits with various tax authorities;

- changes in the valuation of our deferred tax assets and liabilities;
- increases in expenses not deductible for tax purposes, including transaction costs and impairments of goodwill in connection with acquisitions;
- changes in the taxation of share-based compensation;
- changes in tax laws or the interpretation of such tax laws, and changes in generally accepted accounting principles; and
- challenges to the transfer pricing policies related to our structure.

We believe we will likely be classified as a passive foreign investment company for U.S. federal income tax purposes for the current year, which could result in adverse U.S. federal income tax consequences to U.S. investors in our common shares.

Because we do not expect to earn revenue from our business operations during the current taxable year, and because our sole source of income currently is interest on bank accounts held by us, we believe we will likely be classified as a "passive foreign investment company," or PFIC, for the current taxable year. A non-U.S. company will be considered a PFIC for any taxable year if (i) at least 75% of its gross income is passive income (including interest income), or (ii) at least 50% of the value of its assets (based on an average of the quarterly values of the assets during a taxable year) is attributable to assets that produce or are held for the production of passive income. If we are classified as a PFIC in any year with respect to which a U.S. Holder (as defined below under "Material Bermuda and U.S. Federal Income Tax Considerations — Material U.S. Federal Income Tax Considerations to U.S. Holders") owns our common shares, we will continue to be treated as a PFIC with respect to such U.S. Holder in all succeeding years during which the U.S. Holder owns the common shares, regardless of whether we continue to meet the PFIC test described above, unless the U.S. Holder makes or has made a specified election and we cease to be a PFIC. If we are classified as a PFIC for any taxable year during which a U.S. Holder holds our common shares, certain adverse U.S. federal income tax consequences could apply to such U.S. Holder, including (i) the treatment of all or a portion of any gain on disposition as ordinary income, (ii) the application of a deferred interest charge on such gain and the receipt of certain dividends and (iii) the obligation to comply with certain reporting requirements. See "Material Bermuda and U.S. Federal Income Tax Considerations — Material U.S. Federal Income Tax Considerations to U.S. Holders — Passive Foreign Investment Company."

If a U.S. person is treated as owning at least 10% of our common shares, such holder may be subject to adverse U.S. federal income tax consequences.

We believe we are classified as a controlled foreign corporation for the current taxable year and may be classified as a controlled foreign corporation in future taxable years. Even if we were not classified as a controlled foreign corporation, if our group includes one or more U.S. subsidiaries, certain of our non-U.S. subsidiaries could be treated as controlled foreign corporations. If a U.S. Holder (as defined below under "Material Bermuda and U.S. Federal Income Tax Considerations — Material U.S. Federal Income Tax Considerations to U.S. Holders") is treated as owning (directly, indirectly or constructively) at least 10% of the value or voting power of our common shares, such U.S. Holder may be treated as a "United States shareholder" with respect to us (if we are classified as a controlled foreign corporation) and each controlled foreign corporation in our group (if any). A United States shareholder of a controlled foreign corporation may be required to annually report and include in its U.S. taxable income its pro rata share of "Subpart F income," "global intangible low-taxed income" and investments in U.S. property by controlled foreign corporations, regardless of whether we make any distributions. An individual that is a United States shareholder with respect to a controlled foreign corporation generally would not be allowed

certain tax deductions or foreign tax credits that would be allowed to a United States shareholder that is a U.S. corporation. Failure to comply with these reporting obligations may subject you to significant monetary penalties and may prevent the statute of limitations with respect to your U.S. federal income tax return for the year for which reporting was due from starting. We cannot provide any assurances that we will assist investors in determining whether we or any of our non-U.S. subsidiaries, if any, are treated as a controlled foreign corporation or whether such investor is treated as a United States shareholder with respect to any of such controlled foreign corporations. Further, we cannot provide any assurances that we will furnish to any United States shareholders information that may be necessary to comply with the reporting and tax paying obligations discussed above. U.S. Holders should consult their tax advisors regarding the potential application of these rules to their investment in our common shares.

Comprehensive tax reform legislation could adversely affect our business and financial condition.

The U.S. government has recently enacted comprehensive tax legislation that includes significant changes to the taxation of business entities, referenced herein as the Tax Reform Act. These changes include, among others, a permanent reduction to the corporate income tax rate, limiting interest deductions, adopting elements of a territorial tax system and introducing certain anti-base erosion provisions. We continue to examine the impact this tax reform legislation may have on our business. The effect of the Tax Reform Act on our business, whether adverse or favorable, is uncertain, and may not become evident for some period of time. U.S. Holders should consult with their legal and tax advisors regarding any such legislation and the potential tax consequences of investing in our common shares.

SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This prospectus contains forward-looking statements. All statements other than statements of historical facts contained in this prospectus, including statements regarding our future results of operations and financial position, business strategy, prospective products and product candidates, their expected properties, performance and impact on healthcare costs, the expected timeline for achievement of our clinical milestones, the timing of, and potential results from, clinical and other trials, marketing authorization from the FDA or regulatory authorities in other jurisdictions, coverage and reimbursement for procedures using our product candidates, if approved, research and development costs, timing of regulatory filings and feedback, timing and likelihood of success, plans and objectives of management for future operations and future results of anticipated products, are forward-looking statements.

These statements involve known and unknown risks, uncertainties and other important factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements.

In some cases, you can identify forward-looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. The forward-looking statements in this prospectus are only predictions. We have based these forward-looking statements largely on our current expectations and projections about future events and financial trends that we believe may affect our business, financial condition and results of operations. These forward-looking statements speak only as of the date of this prospectus and are subject to a number of risks, uncertainties and assumptions described under the sections in this prospectus entitled "Risk Factors" and "Management's Discussion and Analysis of Financial Condition and Results of Operations" and elsewhere in this prospectus. These forward-looking statements are subject to numerous risks, including, without limitation, the following:

- our status as a development-stage company and our expectation to incur losses in the future;
- our future capital needs and our need to raise additional funds;
- our limited operating history;
- the lengthy and expensive clinical development process with its uncertain outcome and potential for clinical failure or delay;
- the decision by any applicable regulatory authority whether to clear our product candidates for clinical development and, ultimately, whether to approve them for marketing and sale;
- our ability to anticipate and prevent adverse events caused by our product candidates;
- our ability to identify, in-license, acquire, discover or develop additional product candidates;
- our ability to have our product candidates manufactured;
- the market acceptance of our product candidates;
- our ability to timely and successfully develop and commercialize our existing and future product candidates, if approved;
- physician awareness and adoption of our product candidates;
- the size of the market for our product candidates;
- our ability to meet the quality expectations of physicians or patients;
- our ability to improve our product candidates;

- the decision of third-party payors not to cover our product candidates or to require extensive and/or independently performed clinical trials prior to covering or maintaining coverage of our product candidates;
- our ability to successfully manage our growth;
- our ability to avoid product liability claims and maintain adequate product liability insurance;
- our ability to obtain regulatory exclusivity;
- our ability to obtain, maintain, protect and enforce our intellectual property rights related to our product candidates;
- federal, state and foreign regulatory requirements applicable to our product candidates; and
- our ownership concentration may prevent new investors in this offering from influencing significant corporate decisions.

Because forward-looking statements are inherently subject to risks and uncertainties, some of which cannot be predicted or quantified and some of which are beyond our control, you should not rely on these forward-looking statements as predictions of future events. The events and circumstances reflected in our forward-looking statements may not be achieved or occur and actual results could differ materially from those projected in the forward-looking statements. Moreover, we operate in an evolving environment. New risk factors and uncertainties may emerge from time to time, and it is not possible for management to predict all risk factors and uncertainties. As a result of these factors, we cannot assure you that the forward-looking statements in this prospectus will prove to be accurate. Except as required by applicable law, we do not plan to publicly update or revise any forward-looking statements contained herein, whether as a result of any new information, future events, changed circumstances or otherwise.

You should read this prospectus and the documents that we reference in this prospectus and have filed as exhibits to the registration statement, of which this prospectus is a part, completely and with the understanding that our actual future results may be materially different from what we expect. We qualify all of our forward-looking statements by these cautionary statements.

INDUSTRY AND OTHER DATA

Unless otherwise indicated, certain industry data and market data included in this prospectus were obtained from independent third-party surveys, market research, publicly available information, reports of governmental agencies and industry publications and surveys. All of the market data used in this prospectus involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. We believe that the information from these industry publications and surveys included in this prospectus is reliable. The industry in which we operate is subject to a high degree of uncertainty and risk due to a variety of factors, including those described in "Risk Factors" and "Special Note Regarding Forward-Looking Statements" and elsewhere in this prospectus. These and other factors could cause results to differ materially from those expressed in the estimates made by the independent parties and by us.

USE OF PROCEEDS

We estimate that the net proceeds to us from our issuance and sale of 7,000,000 Class A common shares in this offering will be approximately $113.4 million (or $131.0 million if the underwriters exercise in full their option to purchase additional Class A common shares), assuming an initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

Each $1.00 increase (decrease) in the assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the net proceeds to us from this offering by approximately $6.5 million, assuming the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting the estimated underwriting discounts and commissions. Each increase (decrease) of 1,000,000 shares in the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, would increase (decrease) the net proceeds to us from this offering by approximately $16.7 million, assuming no change in the assumed initial public offering price per share and after deducting the estimated underwriting discounts and commissions.

We intend to use the net proceeds from this offering, together with our existing cash resources, as follows:

- approximately $30.0 million to $35.0 million to complete our ongoing Phase 2 proof-of-concept clinical trial of rilonacept for the treatment of pericarditis and, if the results from this trial are favorable, to initiate and advance our planned Phase 3 development of rilonacept;
- approximately $65.0 million to $70.0 million to initiate and advance our planned Phase 2 clinical trial of mavrilimumab for the treatment of GCA;
- approximately $80.0 million to $85.0 million to complete our ongoing Phase 1a/1b clinical trial of KPL-716 in subjects with atopic dermatitis as a proof-of-concept for pruritic conditions and, if the results from the trial are favorable, advance the clinical development of KPL-716 for the treatment of prurigo nodularis and atopic dermatitis; and
- the remainder to fund new and ongoing research and development activities, including to advance the pre-clinical development of KPL-045 and KPL-404, and for working capital and other general corporate activities.

This expected use of net proceeds from this offering represents our current intentions based upon our current plans and business conditions. As of the date of this prospectus, we cannot predict with complete certainty all of the particular uses for the net proceeds from this offering or the actual amounts that we will spend on the uses set forth above. We may also use a portion of the net proceeds to in-license, acquire or invest in additional businesses, technologies, products or assets, although currently we have no specific agreements, commitments or understandings in this regard. The amounts and timing of our actual expenditures will depend on numerous factors, including the progress of our clinical trials, our ability to obtain marketing approval from the FDA for our product candidates and other development and commercialization efforts for our product candidates, as well as the amount of cash used in our operations. We may find it necessary or advisable to use the net proceeds from this offering for other purposes, and as a result, our management will retain broad discretion over the allocation of the net proceeds from this offering.

We anticipate that our existing cash and cash equivalents, together with the anticipated net proceeds from this offering, will be sufficient to fund our operating expenses and capital expenditure requirements into the second quarter of 2020, enabling us (i) to complete our ongoing

clinical trials, (ii) initiate and advance our planned Phase 3 development of rilonacept and our planned Phase 2 clinical trial of mavrilimumab and (iii) advance the clinical development of KPL-716 for the treatment of prurigo nodularis and atopic dermatitis. We have based this estimate on assumptions that may prove to be wrong, and we could utilize our available capital resources sooner than we expect. Following this offering, we will require substantial capital to complete clinical development, seek regulatory approval of, and, if approved, commercialize our product candidates.

Pending the use of the proceeds described above, we plan to invest the net proceeds from this offering in short- and intermediate-term, interest-bearing obligations, investment-grade instruments, certificates of deposit or direct or guaranteed obligations of the U.S. government.

DIVIDEND POLICY

We have never declared or paid any cash dividends on our common shares. In October 2015, we distributed Class B common shares to the then-existing holders of our Class A common shares on a pro rata basis. We intend to retain all of our future earnings, if any, to finance the operation and expansion of our business and do not anticipate paying any cash dividends in the foreseeable future. Any future determination to declare and pay dividends to holders of our common shares will be made at the discretion of our board of directors, which may take into account several factors, including general economic conditions, our financial condition and results of operations, available cash and current and anticipated cash needs, capital requirements, contractual, legal, tax and regulatory restrictions, the implications of the payment of dividends by us to our shareholders and any other factors that our board of directors may deem relevant. In addition, pursuant to the Companies Act, a company may not declare or pay dividends if there are reasonable grounds for believing that (1) the company is, or would after the payment be, unable to pay its liabilities as they become due or (2) that the realizable value of its assets would thereby be less than its liabilities. Under our amended and restated bye-laws, which will be effective immediately following the closing of this offering, each of our common shares is entitled to dividends if, as and when dividends are declared by our board of directors, subject to any preferred dividend right of the holders of any preferred shares.

CAPITALIZATION

The following table sets forth our cash and cash equivalents and our capitalization as of March 31, 2018:

- on an actual basis;
- on a pro forma basis to give effect to:
  - the conversion of all outstanding preferred shares into 5,546,019 Class A common shares, 1,070,502 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares upon the closing of this offering; and
  - the effectiveness of our amended and restated bye-laws immediately following the closing of this offering; and
- on a pro forma as adjusted basis to give further effect to our issuance and sale of 7,000,000 Class A common shares in this offering at an assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

The pro forma and pro forma as adjusted information below is illustrative only, and our capitalization following the closing of this offering will be adjusted based on the actual initial public offering price and other terms of this offering determined at pricing. You should read the following table in conjunction with our consolidated financial statements and the related notes appearing at the end of this prospectus and the sections of the prospectus titled "Selected Consolidated Financial Data," "Management's Discussion and Analysis of Financial Condition and Results of Operations" and "Description of Share Capital."

	As of March 31, 2018		
	Actual	Pro Forma	Pro Forma As Adjusted
	(in thousands, except share and per share data)		
Cash and cash equivalents	$ 221,108	$ 221,108	$ 334,517
Convertible preferred shares (Series A, B and C), par value of $0.000273235 per share; 35,670,093 shares designated, issued and outstanding, actual; and no shares issued and outstanding, pro forma and pro forma as adjusted	$ 310,592	$ —	$ —
Shareholders' equity (deficit):			
Class A common shares, $0.000273235 par value; 5,507,938 shares designated, 724,550 shares issued and outstanding, actual; 6,270,569 shares issued and outstanding, pro forma; 13,270,569 shares issued and outstanding, pro forma as adjusted	—	2	4
Class B common shares, $0.000273235 par value; 3,568,353 shares designated, issued and outstanding, actual; 4,638,855 shares issued and outstanding, pro forma; 4,638,855 shares issued and outstanding, pro forma as adjusted	1	1	1
Class A1 common shares, $0.000273235 par value; no shares designated, issued and outstanding, actual; 12,995,954 shares issued and outstanding, pro forma; 12,995,954 shares issued and outstanding, pro forma as adjusted	—	4	4
Class B1 common shares, $0.000273235 par value; no shares designated, issued and outstanding, actual; 16,057,618 shares issued and outstanding, pro forma; 16,057,618 shares issued and outstanding, pro forma as adjusted	—	4	4
Additional paid-in capital	1,864	312,446	425,824
Accumulated deficit	(106,980)	(106,980)	(106,980)
Total shareholders' equity (deficit)	(105,115)	205,477	318,857
Total capitalization	$ 205,477	$ 205,477	$ 318,857

Each $1.00 increase (decrease) in the assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, additional paid-in capital, total shareholders' equity and total capitalization by $6.5 million, assuming that the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us. Each increase (decrease) of 1,000,000 shares in the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, additional paid-in capital, total shareholders' equity and total capitalization by $16.7 million, assuming no change in the assumed initial public offering price per share and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

The foregoing table excludes:

- 4,702,190 Class A common shares issuable upon exercise of share options outstanding as of March 31, 2018, at a weighted average exercise price of $5.41 per share;
- 4,466,500 Class A common shares that will become available for future issuance under our 2018 Incentive Award Plan, or 2018 Plan, which will become effective in connection with this offering, as well as common shares that become available pursuant to provisions in the 2018 Plan that automatically increase the share reserve under the 2018 Plan as described in "Executive and Director Compensation — Executive Compensation Plans — 2018 Incentive Award Plan" (including 122,971 Class A common shares issuable upon exercise of share options that will be granted to certain of our directors and employees, at an exercise price per share equal to the initial public offering price of our Class A common shares in this offering); and
- 670,000 Class A common shares that will become available for future issuance under our 2018 Employee Share Purchase Plan, or the ESPP, which will become effective in connection with this offering, as well as common shares that become available pursuant to provisions in the ESPP that automatically increase the share reserve under the ESPP as described in "Executive and Director Compensation — Executive Compensation Plans — 2018 Employee Share Purchase Plan".

DILUTION

If you invest in our Class A common shares in this offering, your ownership interest will be immediately diluted to the extent of the difference between the initial public offering price per share and the pro forma as adjusted net tangible book value per common share after this offering.

Our historical net tangible book value (deficit) as of March 31, 2018 was $(106.7) million, or $(24.86) per common share. Our historical net tangible book value (deficit) represents our total tangible assets less our total liabilities and carrying value of our preferred shares, which is not included within our shareholders' equity (deficit). Historical net tangible book value (deficit) per share represents historical net tangible book value (deficit) divided by the 4,292,903 common shares outstanding as of March 31, 2018.

Our pro forma net tangible book value as of March 31, 2018 was $203.9 million, or $5.10 per common share. Pro forma net tangible book value represents the amount of our total tangible assets less our total liabilities, after giving effect to the conversion of all outstanding preferred shares into an aggregate of 35,670,093 common shares upon the closing of this offering. Pro forma net tangible book value per share represents our pro forma net tangible book value divided by the total number of shares outstanding as of March 31, 2018, after giving effect to the pro forma adjustments described above.

After giving further effect to our issuance and sale of 7,000,000 Class A common shares in this offering at an assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, and after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us, our pro forma as adjusted net tangible book value as of March 31, 2018 would have been $318.9 million, or $6.79 per share. This represents an immediate increase in pro forma as adjusted net tangible book value of $1.69 per share to our existing shareholders and immediate dilution of $11.21 per share to new investors purchasing Class A common shares in this offering. Dilution per share to new investors is determined by subtracting the pro forma as adjusted net tangible book value per share after this offering from the initial public offering price per Class A common share paid by new investors. The following table illustrates this dilution on a per share basis:

Assumed initial public offering price per share		$ 18.00
Historical net tangible book value (deficit) per share as of March 31, 2018	$ (24.86)	
Increase per share attributable to the pro forma adjustments described above	29.96	
Pro forma net tangible book value per share as of March 31, 2018	5.10	
Increase in pro forma as adjusted net tangible book value per share attributable to new investors purchasing Class A common shares in this offering	1.69	
Pro forma as adjusted net tangible book value per share after this offering		6.79
Dilution per share to new investors purchasing Class A common shares in this offering		$ 11.21

Each $1.00 increase (decrease) in the assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted net tangible book value per share after this offering by $0.14 and the dilution per share to new investors by $0.86, assuming that the number of Class A common shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting the estimated underwriting discounts and commissions. Each increase of 1,000,000 shares in the number of Class A common shares offered by us would increase our pro forma as adjusted net tangible book value per share after this offering by $0.21 and decrease the dilution per share to new investors by $0.21, assuming no change in the

assumed initial public offering price per share and after deducting the estimated underwriting discounts and commissions. Each decrease of 1,000,000 shares in the number of Class A common shares offered by us would decrease our pro forma as adjusted net tangible book value per share after this offering by $0.22 and increase the dilution per share to new investors by $0.22, assuming no change in the assumed initial public offering price per share and after deducting the estimated underwriting discounts and commissions.

If the underwriters exercise in full their option to purchase additional Class A common shares, the pro forma as adjusted net tangible book value per share after this offering would be $7.01, and the dilution per share to new investors would be $10.99, in each case assuming an initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, and after deducting the estimated underwriting discounts and commissions.

The following table summarizes, on the pro forma as adjusted basis as described above, the total number of common shares purchased from us, the total consideration paid and the average price per share paid or to be paid by existing shareholders and by new investors acquiring our Class A common shares in this offering at an assumed initial public offering price of $18.00 per share, which is the midpoint of the price range set forth on the cover page of this prospectus, before deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us.

	Shares Purchased		Total Consideration		Average Price
	Number	Percent	Amount	Percent	Per Share
Existing shareholders	39,962,996	85.1%	$ 320,028,545	71.8%	$ 8.01
New investors	7,000,000	14.9	126,000,000	28.2	$ 18.00
Total	46,962,996	100.0%	$ 446,028,545	100.0%	

The table above assumes no exercise of the underwriters' option to purchase additional Class A common shares. If the underwriters exercise in full their option to purchase additional Class A common shares, the percentage of our common shares held by existing shareholders would be decreased to 83.2% of the total number of our common shares outstanding after this offering, and the number of shares held by new investors participating in this offering would be increased to 16.8% of the total number of our common shares outstanding after this offering.

The foregoing tables exclude:

- 4,702,190 Class A common shares issuable upon exercise of share options outstanding as of March 31, 2018, at a weighted average exercise price of $5.41 per share;
- 4,466,500 Class A common shares that will become available for future issuance under our 2018 Incentive Award Plan, or 2018 Plan, which will become effective in connection with this offering, as well as common shares that become available pursuant to provisions in the 2018 Plan that automatically increase the share reserve under the 2018 Plan as described in "Executive and Director Compensation — Executive Compensation Plans — 2018 Incentive Award Plan"(including 122,971 Class A common shares issuable upon exercise of share options that will be granted to certain of our directors and employees, at an exercise price per share equal to the initial public offering price of our Class A common shares in this offering); and
- 670,000 Class A common shares that will become available for future issuance under our 2018 Employee Share Purchase Plan, or the ESPP, which will become effective in

connection with this offering, as well as common shares that become available pursuant to provisions in the ESPP that automatically increase the share reserve under the ESPP as described in "Executive and Director Compensation — Executive Compensation Plans — 2018 Employee Share Purchase Plan".

To the extent any of the outstanding share options are exercised, you will experience further dilution as a new investor in this offering. In addition, we may choose to raise additional capital because of market conditions or strategic considerations, even if we believe that we have sufficient funds for our current or future operating plans. If we raise additional capital through the sale of equity or convertible debt securities, the issuance of these securities could result in further dilution to our shareholders.

Certain of our existing shareholders, including shareholders affiliated with one of our directors, have indicated an interest in purchasing an aggregate of approximately $50.0 million of Class A common shares in this offering at the initial public offering price per share. However, because indications of interest are not binding agreements or commitments to purchase, the underwriters may determine to sell more, less or no shares in this offering to any of these existing shareholders, or any of these existing shareholders may determine to purchase more, less or no shares in this offering. The underwriters will receive the same underwriting discount on any shares purchased by these existing shareholders as they will on any other shares sold to the public in this offering.

SELECTED CONSOLIDATED FINANCIAL DATA

You should read the following selected consolidated financial data together with our consolidated financial statements and the related notes appearing at the end of this prospectus and the "Management's Discussion and Analysis of Financial Condition and Results of Operations" section of this prospectus. We have derived the consolidated statement of operations data for the years ended December 31, 2016 and 2017 and the consolidated balance sheet data as of December 31, 2016 and 2017 from our audited consolidated financial statements appearing at the end of this prospectus. We have derived the consolidated statement of operations data for the three months ended March 31, 2017 and 2018 and the consolidated balance sheet data as of March 31, 2018 from our unaudited consolidated financial statements appearing at the end of this prospectus, which have been prepared on the same basis as the audited consolidated financial statements. In the opinion of management, the unaudited data reflects all adjustments, consisting only of normal recurring adjustments, necessary for a fair statement of the financial information in those statements. Our historical results are not necessarily indicative of results that may be expected in any future period, and our results for any interim period are not necessarily indicative of results that may be expected for any full year.

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
	(in thousands, except share and per share data)			
Consolidated Statement of Operations Data:				
Operating expenses:				
Research and development	$ 17,439	$ 56,357	$ 3,145	$ 12,630
General and administrative	6,563	9,043	1,903	3,710
Total operating expenses	24,002	65,400	5,048	16,340
Loss from operations	(24,002)	(65,400)	(5,048)	(16,340)
Interest income	65	529	74	305
Loss before benefit (provision) for income taxes	(23,937)	(64,871)	(4,974)	(16,035)
Benefit (provision) for income taxes	(36)	(2)	33	53
Net loss	$ (23,973)	$ (64,873)	$ (4,941)	$ (15,982)
Net loss per share attributable to common shareholders—basic and diluted[1]	$ (91.61)	$ (35.85)	$ (3.52)	$ (6.45)
Weighted average common shares outstanding—basic and diluted[1]	261,695	1,809,751	1,405,400	2,478,903
Pro forma net loss per share attributable to common shareholders—basic and diluted (unaudited)[1]		$ (2.74)		$ (0.49)
Pro forma weighted average common shares outstanding—basic and diluted (unaudited)[1]		23,638,410		32,466,951

(1) See Note 11 to our consolidated financial statements included elsewhere in this prospectus for further details on the calculation of basic and diluted net loss per share attributable to common shareholders and on the calculation of pro forma basic and diluted net loss per share attributable to common shareholders.

	As of December 31,		As of March 31,
	2016	2017	2018
	(in thousands)		
Consolidated Balance Sheet Data:			
Cash and cash equivalents	$ 55,970	$ 45,555	$ 221,108
Working capital[(1)]	54,032	29,674	203,185
Total assets	56,467	47,492	224,775
Convertible preferred shares	79,897	119,770	310,592
Total shareholders' deficit	(25,732)	(89,708)	(105,115)

(1) We define working capital as current assets less current liabilities.

MANAGEMENT'S DISCUSSION AND ANALYSIS OF FINANCIAL CONDITION AND RESULTS OF OPERATIONS

You should read the following discussion and analysis of our financial condition and results of operations together with our consolidated financial statements and the related notes and the other financial information included elsewhere in this prospectus. Some of the information contained in this discussion and analysis or set forth elsewhere in this prospectus, including information with respect to our plans and strategy for our business and related financing, includes forward-looking statements that involve risks and uncertainties. As a result of many factors, including those factors set forth in the "Risk Factors" section of this prospectus, our actual results could differ materially from the results described in or implied by these forward-looking statements.

Overview

We are a clinical-stage biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have a pipeline of product candidates across various stages of development, currently focused on autoinflammatory and autoimmune conditions. We have three clinical-stage product candidates, one of which is anticipated to commence a Phase 3 clinical trial in 2018. We follow a disciplined and methodical approach to selectively identify and acquire product candidates with strong biologic rationales or validated mechanisms of action. We believe that each of our product candidates has the potential to address multiple indications.

Since our inception in 2015, we have devoted substantially all of our efforts and financial resources to organizing and staffing our company, business planning, raising capital, acquiring, in-licensing or discovering product candidates and securing related intellectual property rights and conducting research and development activities for our programs. We do not have any products approved for sale and have not generated any revenue from product sales. We have funded our operations to date primarily with proceeds from the sale of preferred shares. Through March 31, 2018, we had received net proceeds of $310.6 million from the sale of preferred shares.

We have incurred significant operating losses since inception. Our ability to generate product revenue sufficient to achieve profitability will depend heavily on the successful development and eventual commercialization of one or more of our current or future product candidates. Our net losses were $24.0 million and $64.9 million for the years ended December 31, 2016 and 2017, respectively and were $4.9 million and $16.0 million for the three months ended March 31, 2017 and 2018, respectively. As of December 31, 2017 and March 31, 2018, we had an accumulated deficit of $91.0 million and $107.0 million, respectively. We expect to continue to incur significant operating losses for at least the next several years as we advance our product candidates through all stages of development and clinical trials and, ultimately, seek regulatory approval. In addition, if we obtain marketing approval for any of our product candidates, we expect to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates. Furthermore, upon the closing of this offering, we expect to incur additional costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses that we did not incur as a private company.

As a result, we will need substantial additional funding to support our continuing operations and pursue our growth strategy. Until such time as we can generate significant revenue from product sales, if ever, we expect to finance our operations through the sale of equity, debt financings or other capital sources, which may include collaborations with other companies or other strategic transactions. We may be unable to raise additional funds or enter into such other agreements or arrangements when needed on favorable terms, or at all. If we fail to raise capital or

enter into such agreements as and when needed, we may have to significantly delay, scale back or discontinue the development and commercialization of one or more of our product candidates or delay our pursuit of potential in-licenses or acquisitions.

Because of the numerous risks and uncertainties associated with product development, we are unable to predict the timing or amount of increased expenses or when or if we will be able to achieve or maintain profitability. Even if we are able to generate product sales, we may not become profitable. If we fail to become profitable or are unable to sustain profitability on a continuing basis, then we may be unable to continue our operations at planned levels and be forced to reduce or terminate our operations.

As of December 31, 2017 and March 31, 2018, we had cash and cash equivalents of $45.6 million and $221.1 million, respectively. We believe that our existing cash and cash equivalents, will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months. We have based this estimate on assumptions that may prove to be wrong, and we could exhaust our available capital resources sooner than we expect. See "— Liquidity and Capital Resources."

Components of Our Results of Operations

Revenue

To date, we have not generated any revenue from product sales and do not expect to generate any revenue from the sale of products in the foreseeable future. If our development efforts for our product candidates are successful and result in regulatory approval, we may generate revenue in the future from product sales.

Operating Expenses

Research and Development Expenses

Research and development expenses consist primarily of costs incurred in connection with the discovery and development of our product candidates. We expense research and development costs as incurred. These expenses may include:

- expenses incurred to conduct the necessary pre-clinical studies and clinical trials required to obtain regulatory approval;
- expenses incurred under agreements with CROs, that are primarily engaged in the oversight and conduct of our clinical trials and CMOs, that are primarily engaged to provide pre-clinical and clinical drug substance and product for our research and development programs;
- other costs related to acquiring and manufacturing pre-clinical studies and clinical trial materials, including manufacturing validation batches, as well as investigative sites and consultants that conduct our clinical trials, pre-clinical studies and other scientific development services;
- payments made in cash or equity securities under third-party licensing, acquisition and option agreements;
- employee-related expenses, including salaries and benefits, travel and share-based compensation expense for employees engaged in research and development functions;
- costs related to compliance with regulatory requirements; and
- allocated facilities-related costs, depreciation and other expenses, which include rent and utilities.

We recognize external development costs based on an evaluation of the progress to completion of specific tasks using information provided to us by our service providers. This process involves reviewing open contracts and purchase orders, communicating with our personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of actual costs. Nonrefundable advance payments for goods or services to be received in the future for use in research and development activities are recorded as prepaid expenses. Such amounts are recognized as an expense as the goods are delivered or the related services are performed, or until it is no longer expected that the goods will be delivered or the services rendered.

Our direct research and development expenses are tracked on a program-by-program basis for our product candidates and consist primarily of external costs, such as fees paid to outside consultants, CROs, CMOs and research laboratories in connection with our pre-clinical development, process development, manufacturing and clinical development activities. Our direct research and development expenses by program also include fees incurred under license, acquisition and option agreements. We do not allocate employee costs or facility expenses, including depreciation or other indirect costs, to specific programs because these costs are deployed across multiple programs and, as such, are not separately classified. We use internal resources primarily to conduct our research and discovery as well as for managing our pre-clinical development, process development, manufacturing and clinical development activities.

The table below summarizes our research and development expenses incurred by program:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
	(in thousands)			
Rilonacept[1]	$ —	$ 6,301	$ —	$ 1,223
Mavrilimumab[2]	—	18,000	—	278
KPL-716[3]	14,870	24,164	2,201	7,215
KPL-045[4]	—	1,654	—	437
KPL-404[5]	—	549	—	374
Unallocated research and development expenses	2,569	5,689	944	3,103
Total research and development expenses	$ 17,439	$ 56,357	$ 3,145	$ 12,630

(1) The amount for the year ended December 31, 2017 includes expense of $5.0 million related to an upfront payment under our license agreement with Regeneron.

(2) The amount for the year ended December 31, 2017 consists of expense of $18.0 million related to an upfront payment and an accrued milestone under our license agreement with MedImmune.

(3) The amount for the year ended December 31, 2016 includes expense of $11.5 million related to an upfront payment and $0.5 million related to a technology transfer payment under our asset purchase agreement with Biogen. The amount for the year ended December 31, 2017 includes expense of $4.0 million related to a milestone payment under our asset purchase agreement with Biogen associated with the achievement of a specified clinical milestone event.

(4) The amount for the year ended December 31, 2017 includes expense of $1.5 million related to an upfront payment under our license agreement with Novo Nordisk. The amount for the three months ended March 31, 2018 includes expense of $0.2 million related to a technology transfer payment under our license agreement with Novo Nordisk.

(5) The amount for the year ended December 31, 2017 includes expense of $0.5 million related to upfront payments for the initial option period under our stock purchase option agreement with Primatope.

Research and development activities are central to our business model. Product candidates in later stages of clinical development generally have higher development costs than those in earlier stages of clinical development, primarily due to the increased size and duration of later-stage clinical trials. As a result, we expect that our research and development expenses will increase substantially over the next several years as we complete our ongoing and planned clinical trials for rilonacept, mavrilimumab and KPL-716, as well as conduct other pre-clinical and clinical development including regulatory filings for our other product candidates and our discovery research efforts and increase personnel costs, including costs associated with share-based compensation. We also expect to incur additional expenses related to milestone and royalty payments payable to third parties with whom we have entered into license, acquisition and option agreements to acquire the rights to our product candidates.

The successful development and commercialization of our product candidates is highly uncertain. At this time, we cannot reasonably estimate or know the nature, timing and costs of the efforts that will be necessary to complete the pre-clinical and clinical development of any of our product candidates or when, if ever, material net cash inflows may commence from any of our product candidates. This uncertainty is due to the numerous risks and uncertainties associated with product development and commercialization, including the uncertainty of:

- the scope, progress, outcome and costs of our pre-clinical development activities, clinical trials and other research and development activities;
- establishing an appropriate safety and efficacy profile with IND-enabling and clinical studies;
- successful patient enrollment in and the initiation and completion of clinical trials;
- the timing, receipt and terms of any marketing approvals from applicable regulatory authorities including the FDA;
- the extent of any required post-marketing approval commitments to applicable regulatory authorities;
- establishing clinical and commercial manufacturing capabilities or making arrangements with third-party manufacturers in order to ensure that we or our third-party manufacturers are able to make product successfully;
- development and timely delivery of clinical-grade and commercial-grade drug formulations that can be used in our clinical trials and for commercial launch;
- obtaining, maintaining, defending and enforcing patent claims and other intellectual property rights;
- significant and changing government regulation;
- launching commercial sales of our product candidates, if and when approved, whether alone or in collaboration with others; and
- maintaining a continued acceptable safety profile of the product candidates following approval if any of our product candidates are approved.

General and Administrative Expenses

General and administrative expenses consist primarily of salaries and benefits, travel and share-based compensation expense for personnel in executive, business development, finance, human resources, legal and support personnel functions. General and administrative expenses also include insurance and professional fees for legal, patent, consulting, accounting and audit services.

We expect that our general and administrative expenses will increase in the future as we increase our headcount to support our continued research activities and development of our product candidates. We also anticipate that we will incur increased accounting, audit, legal, regulatory, compliance and director and officer insurance costs as well as investor and public relations expenses associated with being a public company. Additionally, if and when we believe a regulatory approval of a product candidate appears likely, we anticipate an increase in payroll and expense as a result of our preparation for commercial operations, especially as it relates to the sales and marketing of our product candidate.

Interest Income

Interest income consists of income recognized from investments in money market funds and U.S. Treasury securities.

Income Taxes

As a company incorporated in Bermuda, we are principally subject to taxation in Bermuda. Under the current laws of Bermuda, tax on a company's income is assessed at a zero percent tax rate. As a result, we have not recorded any income tax benefits from our losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards will be available to us for those losses. Our provision for income taxes relates to taxable income generated by our wholly owned U.S. subsidiary, Kiniksa Pharmaceuticals Corp., under a cost-plus arrangement with our parent company, Kiniksa Pharmaceuticals, Ltd. Kiniksa Pharmaceuticals Corp. is subject to federal and state income taxes in the United States.

As of December 31, 2017, we had state research and development tax credit carryforwards of approximately $0.1 million available to reduce future tax liabilities, which begin to expire in 2031 through 2032.

Results of Operations

Comparison of the Three Months Ended March 31, 2017 and 2018

The following table summarizes our results of operations for the three months ended March 31, 2017 and 2018:

	Three Months Ended March 31,		
	2017	2018	Change
		(in thousands)	
Operating expenses:			
Research and development	$ 3,145	$ 12,630	$ 9,485
General and administrative	1,903	3,710	1,807
Total operating expenses	5,048	16,340	11,292
Loss from operations	(5,048)	(16,340)	(11,292)
Interest income	74	305	231
Loss before provision for income taxes	(4,974)	(16,035)	(11,061)
Benefit (provision) for income taxes	33	53	20
Net loss	$ (4,941)	$ (15,982)	$ (11,041)

Research and Development Expenses

	Three Months Ended March 31,		
	2017	2018	Change
		(in thousands)	
Direct research and development expenses by program:			
Rilonacept	$ —	$ 1,223	$ 1,223
Mavrilimumab	—	278	278
KPL-716	2,201	7,215	5,014
KPL-045	—	437	437
KPL-404	—	374	374
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	738	2,457	1,719
Other	206	646	440
Total research and development expenses	$ 3,145	$ 12,630	$ 9,485

Research and development expenses were $12.6 million for the three months ended March 31, 2018, compared to $3.1 million for the three months ended March 31, 2017. The increase of $9.5 million was primarily due to an increase in external fees related to our development programs, of which there were five in 2018 and one in 2017, as well as an increase of $2.2 million in unallocated research and development expenses.

The direct costs of $1.2 million for our rilonacept program during the three months ended March 31, 2018 were due to expenses related to clinical research and development with our open-label Phase 2 proof-of-concept clinical trial. We had no direct costs for our rilonacept program during the three months ended March 31, 2017.

The direct costs of $0.3 million for our mavrilimumab program during the three months ended March 31, 2018 were due to expenses related primarily to preparation for our planned Phase 2 clinical trial. We had no direct costs for our mavrilimumab program during the three months ended March 31, 2017.

The direct costs of our KPL-716 program were $7.2 million during the three months ended March 31, 2018, compared to $2.2 million during the three months ended March 31, 2017. The increase in $5.0 million in direct costs for our KPL-716 program during the three months ended March 31, 2018 was primarily due to increased expenses related to our Phase 1a/1b clinical trial and our LOTUS-PN observational study including manufacturing and development costs related to clinical drug supply, partially offset by a decrease in other research and development studies related to this program.

The direct costs of $0.4 million for our KPL-045 program during the three months ended March 31, 2018 were due to $0.2 million of direct costs related to clinical research and development as well as a $0.2 million payment related to technology transfer under our agreement with Novo Nordisk. We had no direct costs for our KPL-045 program during the three months ended March 31, 2017.

The direct costs of $0.4 million for our KPL-404 program during the three months ended March 31, 2018 were due to expenses related to clinical research and development, including manufacturing development. We had no direct costs for our KPL-404 program during the three months ended March 31, 2017.

Unallocated research and development expenses were $3.1 million for the three months ended March 31, 2018 compared to $0.9 million for the three months ended March 31, 2017. The increase of $2.2 million in unallocated research and development expenses was due to an increase

of $1.7 million in personnel-related costs, including share-based compensation, and an increase of $0.4 million in other costs. The increase in personnel-related costs was primarily due to the hiring of additional personnel in our research and development functions, particularly those responsible for coordinating with CMOs on development and manufacturing of drug supply and coordinating with CROs on the conduct and oversight of our current and planned clinical trials as well as research studies and development programs for our product candidates. Personnel-related costs for the three months ended March 31, 2018 and 2017 included share-based compensation of $0.2 million and $29,700, respectively. The increase in other costs was primarily due to a $0.2 million increase in professional fees, a $0.1 million increase in travel expenses and a $0.1 million increase in certain allocated facilities-related costs.

General and Administrative Expenses

General and administrative expenses were $3.7 million for the three months ended March 31, 2018 compared to $1.9 million for the three months ended March 31, 2017. The increase of $1.8 million was due to increases of $0.7 million in personnel-related costs and $1.1 million in professional fees. The increase in personnel-related costs was due to the hiring of additional personnel in our general and administrative functions, primarily in our corporate, finance and human resources departments, as we continued to expand our operations to support the organization. Personnel-related costs for the three months ended March 31, 2018 and 2017 included share-based compensation of $0.4 million and $0.1 million, respectively. Professional fees increased due to legal costs incurred in connection with maintaining and registering worldwide patents and costs associated with our ongoing business operations, as well as higher accounting, recruiting and market research expenses.

Interest Income

Interest income was $0.3 million for the three months ended March 31, 2018 compared to $0.1 million for the three months ended March 31, 2017. The increase was due to both higher average invested cash balances and higher interest rates on U.S. Treasury securities in 2018.

Benefit (Provision) for Income Taxes

We recorded an insignificant benefit for income taxes for the three months ended March 31, 2018 and 2017.

Comparison of the Years Ended December 31, 2016 and 2017

The following table summarizes our results of operations for the years ended December 31, 2016 and 2017:

	Year Ended December 31,		
	2016	2017	Change
	(in thousands)		
Operating expenses:			
Research and development	$ 17,439	$ 56,357	$ 38,918
General and administrative	6,563	9,043	2,480
Total operating expenses	24,002	65,400	41,398
Loss from operations	(24,002)	(65,400)	(41,398)
Interest income	65	529	464
Loss before provision for income taxes	(23,937)	(64,871)	(40,934)
Provision for income taxes	(36)	(2)	34
Net loss	$ (23,973)	$ (64,873)	$ (40,900)

Research and Development Expenses

	Year Ended December 31,		
	2016	2017	Change
	(in thousands)		
Direct research and development expenses by program:			
Rilonacept	$ —	$ 6,301	$ 6,301
Mavrilimumab	—	18,000	18,000
KPL-716	14,870	24,164	9,294
KPL-045	—	1,654	1,654
KPL-404	—	549	549
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	1,837	4,576	2,739
Other	732	1,113	381
Total research and development expenses	$ 17,439	$ 56,357	$ 38,918

Research and development expenses were $56.4 million for the year ended December 31, 2017, compared to $17.4 million for the year ended December 31, 2016. The increase of $38.9 million was primarily due to an increase in external fees related to our development programs as well as an increase of $3.1 million in unallocated research and development expenses.

The direct costs of $6.3 million for our rilonacept program during the year ended December 31, 2017 were due to a $5.0 million upfront payment made under our license agreement with Regeneron, as well as $1.3 million of clinical research and development costs associated with the commencement of our open-label Phase 2 proof-of-concept clinical trial. We had no direct costs for our rilonacept program during the year ended December 31, 2016.

The direct costs of $18.0 million for our mavrilimumab program during the year ended December 31, 2017 were due to an $8.0 million upfront payment made under our license agreement with MedImmune as well as an accrued milestone of $10.0 million, as we have

determined the payment related to the milestone to be probable. We had no direct costs for our mavrilimumab program during the year ended December 31, 2016.

The direct costs for our KPL-716 program were $24.2 million during the year ended December 31, 2017, compared to $14.9 million during the year ended December 31, 2016. The increase of $9.3 million in direct costs for our KPL-716 program during the year ended December 31, 2017 was primarily due to expenses related to our Phase 1a/1b clinical trial, including a $4.0 million milestone payment made to Biogen upon the achievement of a specified clinical milestone event, as well as expenses related to our LOTUS-PN observational study, manufacturing development costs for clinical drug supply and other research and development studies. During the year ended December 31, 2016, direct costs for our KPL-716 program included expenses of $11.5 million related to an upfront payment and $0.5 million related to a technology transfer payment, each under our agreement with Biogen.

The direct costs of $1.7 million for our KPL-045 program during the year ended December 31, 2017 were primarily due to a $1.5 million upfront payment made under our license agreement with Novo Nordisk. We had no direct costs for our KPL-045 program during the year ended December 31, 2016.

The direct costs of $0.5 million for our KPL-404 program during the year ended December 31, 2017 were due to $0.5 million of upfront payments made in connection with the initial option period under our stock purchase option agreement with Primatope. We had no direct costs for our KPL-404 program during the year ended December 31, 2016.

Unallocated research and development expenses were $5.7 million for the year ended December 31, 2017, compared to $2.6 million for the year ended December 31, 2016. The increase of $3.1 million in unallocated research and development expenses was due to an increase of $2.7 million in personnel-related costs, including share-based compensation, and an increase of $0.4 million in other costs. The increase in personnel-related costs was primarily due to the hiring of additional personnel in our research and development functions, particularly those responsible for coordinating with CMOs on development and manufacturing of drug supply for our product candidates and coordinating with CROs on the conduct and oversight of our Phase 1a/1b clinical trial and LOTUS-PN observational study for our KPL-716 program and our open-label Phase 2 proof-of-concept clinical trial for our rilonacept program. Personnel-related costs for the years ended December 31, 2017 and 2016 included share-based compensation of $0.3 million and $0.1 million, respectively. The increase in other costs was primarily due to a $0.2 million increase in travel expense and a $0.1 million increase in certain allocated facilities-related costs and information technology expenses.

General and Administrative Expenses

General and administrative expenses were $9.0 million for the year ended December 31, 2017, compared to $6.6 million for the year ended December 31, 2016. The increase of $2.5 million was primarily due to increases of $1.4 million in personnel-related costs and $1.1 million in professional fees. The increase in personnel-related costs was due to the hiring of additional personnel in our general and administrative functions, primarily in our legal and finance departments, as we continued to expand our operations to support the organization. Personnel-related costs for the years ended December 31, 2017 and 2016 included share-based compensation of $0.6 million and $0.3 million, respectively. Professional fees increased due to legal costs incurred in connection with maintaining and registering worldwide patents and costs associated with our ongoing business operations, as well higher accounting, consulting and market research expenses.

Interest Income

Interest income was $0.5 million for the year ended December 31, 2017, compared to $0.1 million for the year ended December 31, 2016. The increase was due to both higher average invested cash balances and higher interest rates on U.S. Treasury securities in 2017.

Provision for Income Taxes

We recorded an insignificant provision for income taxes for the years ended December 31, 2017 and 2016.

Liquidity and Capital Resources

Since our inception, we have not generated any revenue from any sources, including from product sales, and have incurred significant operating losses and negative cash flows from our operations. We have funded our operations to date primarily with proceeds from the sale of preferred shares. Through March 31, 2018, we had received net proceeds of $310.6 million from sales of our preferred shares. As of December 31, 2017 and March 31, 2018, we had cash and cash equivalents of $45.6 million and $221.1 million, respectively.

Cash Flows

The following table summarizes our cash flows for each of the periods presented:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
	(in thousands)			
Net cash used in operating activities	$ (21,867)	$ (50,219)	$ (5,679)	$ (14,972)
Net cash used in investing activities	(3)	(69)	(18)	(75)
Net cash provided by financing activities	42,509	39,873	39,873	190,810
Net increase (decrease) in cash and cash equivalents and restricted cash	$ 20,639	$ (10,415)	$ 34,176	$ 175,763

Operating Activities

During the three months ended March 31, 2018, operating activities used $15.0 million of cash, primarily resulting from our net loss of $16.0 million, partially offset by non-cash charges of $0.5 million and net cash provided by changes in our operating assets and liabilities of $0.5 million. Net cash provided by changes in our operating assets and liabilities for the three months ended March 31, 2018 consisted of a $1.1 million increase in accounts payable and a $0.2 million decrease in prepaid expenses and other current assets, both partially offset by a $0.8 million decrease in accrued expenses. The increase in accounts payable was due to our increased level of operating activities and the timing of vendor invoicing and payments. The decrease in prepaid expenses and other current assets was primarily due to a decrease in prepaid expenses to CMOs related to manufacturing development related to our product candidates. The decrease in accrued expenses was primarily due to a decrease in accrued employee compensation expense related to the payment of the 2017 annual bonus during the three months ended March 31, 2018.

During the three months ended March 31, 2017, operating activities used $5.7 million of cash, primarily resulting from our net loss of $4.9 million and $0.8 million in net cash used in changes in our operating assets and liabilities, partially offset by non-cash charges of $0.1 million. Net cash used in changes in our operating assets and liabilities for the three months ended March 31, 2017

consisted of a $1.1 million decrease in accrued expenses, partially offset by a $0.4 million increase in accounts payable. The decrease in accrued expenses was primarily due to the payment of the 2016 annual bonus during the three months ended March 31, 2017. The increase in accounts payable was due to our increased level of operating activities and the timing of vendor invoicing and payments.

During the year ended December 31, 2017, operating activities used $50.2 million of cash, primarily resulting from our net loss of $64.9 million, partially offset by non-cash charges of $0.7 million and net cash provided by changes in our operating assets and liabilities of $13.9 million. Net cash provided by changes in our operating assets and liabilities for the year ended December 31, 2017 consisted of a $14.1 million increase in accrued expenses and a $1.0 million increase in accounts payable, both partially offset by a $1.2 million increase in prepaid expenses and other current assets. The increase in accrued expenses was primarily due to an accrued milestone of $10.0 million related to our mavrilimumab program, increased clinical trial and manufacturing activities as well as increased accrued legal and professional fees and accrued employee compensation-related expenses. The increase in accounts payable was due to our increased level of operating activities and the timing of vendor invoicing and payments. The increase in prepaid expenses and other current assets was primarily due to prepaid clinical trial and manufacturing costs associated with our research and development programs.

During the year ended December 31, 2016, operating activities used $21.9 million of cash, primarily resulting from our net loss of $24.0 million, partially offset by non-cash charges of $0.3 million and net cash provided by changes in our operating assets and liabilities of $1.8 million. Net cash provided by changes in our operating assets and liabilities for the year ended December 31, 2016 consisted primarily of a $1.9 million increase in accrued expenses, partially offset by a $0.2 million increase in prepaid expenses and other current assets. The increase in accrued expenses was primarily due to increased research and development costs, an accrued expense related to technology transfer, regulatory consulting costs and accrued compensation expense. The increase in prepaid expenses and other current assets was primarily due to prepaid manufacturing costs and recording an income tax receivable.

Investing Activities

During the three months ended March 31, 2018, investing activities used $0.1 million of cash, consisting of purchases of property and equipment.

During the three months ended March 31, 2017, cash used in investing activities was not significant.

During the year ended December 31, 2017, investing activities used $0.1 million of cash, consisting of purchases of property and equipment.

During the year ended December 31, 2016, we used an insignificant amount of cash in investing activities, consisting of purchases of property and equipment.

Financing Activities

During the three months ended March 31, 2018, net cash provided by financing activities was $190.8 million, primarily consisting of net proceeds from our issuance and sale of Series C preferred shares.

During the three months ended March 31, 2017, net cash provided by financing activities was $39.9 million, consisting of net proceeds from our issuance and sale of Series B preferred shares.

During the year ended December 31, 2017, net cash provided by financing activities was $39.9 million, consisting of net proceeds from our issuance and sale of Series B preferred shares.

During the year ended December 31, 2016, net cash provided by financing activities was $42.5 million, consisting of net proceeds from our issuance and sale of Series A preferred shares.

Funding Requirements

We expect our expenses to increase substantially in connection with our ongoing activities, particularly as we advance the pre-clinical activities and clinical trials of our product candidates. In addition, upon the closing of this offering, we expect to incur additional costs associated with operating as a public company. Our expenses will also increase as we:

- continue to conduct our current clinical trials and initiate our planned clinical trials of rilonacept, mavrilimumab and KPL-716;
- advance pre-clinical development of our early-stage programs, KPL-045 and KPL-404;
- manufacture, or have manufactured on our behalf, our pre-clinical and clinical drug material and develop processes for late state and commercial manufacturing;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- establish a sales, marketing, medical affairs and distribution infrastructure to commercialize any product candidates for which we may obtain marketing approval and intend to commercialize on our own;
- hire additional clinical, quality control and scientific personnel;
- expand our operational, financial and management systems and increase personnel, including personnel to support our clinical development, manufacturing and commercialization efforts and our operations as a public company;
- maintain, expand and protect our intellectual property portfolio; and
- acquire or in-license other product candidates and technologies.

We believe that our existing cash and cash equivalents, will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months. We have based these estimates on assumptions that may prove to be wrong, and we could utilize our available capital resources sooner than we expect. We anticipate that we may require additional capital if we choose to pursue in-licenses or acquisitions of other product candidates. If we receive regulatory approval for rilonacept or our other product candidates, we expect to incur significant commercialization expenses related to product manufacturing, sales, marketing and distribution, depending on where we choose to commercialize.

Because of the numerous risks and uncertainties associated with research, development and commercialization of biologic product candidates, we are unable to estimate the exact amount of our working capital requirements. Our future funding requirements will depend on and could increase significantly as a result of many factors, including:

- the scope, progress, results and costs of researching and developing our product candidates, and conducting pre-clinical and clinical trials;
- the costs, timing and outcome of regulatory review of our product candidates;
- the costs, timing and ability to manufacture our product candidates to supply our clinical and pre-clinical development efforts and our clinical trials;

- the costs of future activities, including product sales, medical affairs, marketing, manufacturing and distribution, for any of our product candidates for which we receive marketing approval;
- the costs of manufacturing commercial-grade product and necessary inventory to support commercial launch;
- the ability to receive additional non-dilutive funding, including grants from organizations and foundations;
- the revenue, if any, received from commercial sale of our products, should any of our product candidates receive marketing approval;
- the costs of preparing, filing and prosecuting patent applications, maintaining and enforcing our intellectual property rights and defending intellectual property-related claims;
- our ability to establish and maintain collaborations on favorable terms, if at all;
- the extent to which we acquire or in-license other product candidates and technologies; and
- the timing, receipt and amount of sales of, or milestone payments related to or royalties on, our current or future product candidates, if any.

Until such time, if ever, as we can generate substantial product revenue, we expect to finance our cash needs through a combination of public or private equity offerings, debt financings, governmental funding, collaborations, strategic partnerships or marketing, distribution or licensing arrangements with third parties. To the extent that we raise additional capital through the sale of equity or convertible debt securities, your ownership interest may be materially diluted, and the terms of such securities could include liquidation or other preferences that adversely affect your rights as a common shareholder. Debt financing and preferred equity financing, if available, may involve agreements that include restrictive covenants that limit our ability to take specified actions, such as incurring additional debt, making capital expenditures or declaring dividends. In addition, debt financing would result in fixed payment obligations.

If we raise funds through governmental funding, collaborations, strategic partnerships or marketing, distribution or licensing arrangements with third parties, we may have to relinquish valuable rights to our technologies, future revenue streams, research programs or product candidates or grant licenses on terms that may not be favorable to us. If we are unable to raise additional funds when needed, we may be required to delay, reduce or eliminate our product development or future commercialization efforts.

Contractual Obligations and Commitments

The following table summarizes our contractual obligations as of December 31, 2017 and the effects that such obligations are expected to have on our liquidity and cash flows in future periods:

	Payments Due by Period				
	Less than 1 Year	1 to 3 Years	4 to 5 Years	More than 5 Years	Total
	(in thousands)				
Accrued milestone[1]	$ 10,000	$ —	$ —	$ —	$ 10,000
Manufacturing commitments[2]	7,766	—	—	—	7,766
Operating lease commitments[3]	270	—	—	—	270
Total	$ 18,036	$ —	$ —	$ —	$ 18,036

(1) Represents a payment of $10.0 million we are obligated to make under our license agreement with MedImmune upon the earlier to occur of (a) the first achievement of a specified regulatory milestone for a product licensed under the agreement and (b) December 31, 2018.

(2) Amounts in the table reflect commitments for costs associated with our external CMOs, which we have engaged to manufacture pre-clinical and clinical trial materials. Manufacturing commitments include agreements that are enforceable and legally binding on us and that specify all significant terms, including fixed or minimum quantities to be purchased; fixed, minimum or variable price provisions; and the approximate timing of the transaction. For obligations with cancellation provisions, the amounts included in the preceding table are limited to the non-cancelable portion of the agreement terms or the minimum cancellation fee. As of March 31, 2018, non-cancelable commitments associated with our external CMOs were $5.9 million.

(3) Represents minimum payments due for the lease of office space by our wholly owned U.S. subsidiary, Kiniksa Pharmaceuticals Corp., or Kiniksa US, in Wellesley Hills, Massachusetts under an operating lease agreement that expires in August 2018. In March 2018, Kiniksa US entered into a lease agreement for our new U.S. headquarters, which expires in July 2021. Upon execution of the lease, we made a prepayment of $0.1 million for August 2018 rent. The lease requires monthly rental payments of $0.1 million beginning in September 2018. The lease requires future rental payments of $0.3 million during the year ending December 31, 2018, an aggregate of $1.6 million during the years ending December 31, 2019 and 2020 and $0.5 million during the year ending December 31, 2021. Such amounts are not reflected in the table.

Our contracts with CMOs, CROs and other third parties for the manufacture of our product candidates and to support clinical trials and pre-clinical research studies and testing are generally cancelable by us upon prior notice. Payments due upon cancellation consisting only of payments for services provided or expenses incurred, including noncancelable obligations of our service providers, up to the date of cancellation are not included in the preceding table as the amount and timing of such payments are not known.

Under various agreements with third parties, we have agreed to make milestone payments, pay royalties, annual maintenance fees and to meet due diligence requirements based upon specified milestones. We generally have not included any contingent payment obligations, such as milestones, royalties or due diligence, in the table above as the amount, timing and likelihood of such payments are not known. We have not included any of the annual maintenance fee payments in the above table, as although the amount and timing are known, we cannot currently determine the final termination dates of the agreements and, as a result, we cannot determine the total amounts of such payments we will be required to make under the agreements.

Under our license agreement with Regeneron, we are obligated to make future regulatory milestone payments of $27.5 million in the aggregate. Thereafter, we have agreed to evenly split profits on our sales of rilonacept with Regeneron after deducting certain commercialization expenses subject to specified limits.

Under our license agreement with MedImmune, we are obligated to make future clinical, regulatory and initial sales milestone payments of up to $72.5 million in aggregate for the first two indications we develop, including a milestone payment of $10.0 million upon the earlier to occur of

a specified regulatory milestone and December 31, 2018, and clinical and regulatory milestone payments of up to $15.0 million in the aggregate for each subsequent indication. The $10.0 million milestone payment was accrued on our consolidated balance sheet as of December 31, 2017 and recognized as research and development expense during the year ended December 31, 2017. Such payment is included in the table above. We are also obligated to make milestone payments to MedImmune of up to $85.0 million upon the achievement of annual net sales thresholds of up to, but excluding, $1.0 billion in annual net sales as well as additional milestone payments aggregating up to $1.1 billion upon the achievement of additional specified annual net sales thresholds starting at $1.0 billion and higher. Commencing on the first commercial sale of licensed products, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. We must pay such royalties on a product-by-product and country-by-country basis until the latest to occur of the expiration of licensed patents, the expiration of regulatory exclusivity or the tenth anniversary of first commercial sale of such product in such country.

Under our asset purchase agreement with Biogen, we are obligated to make future milestone payments of up to $325.0 million upon the achievement of specified clinical and regulatory milestones as well as upon the achievement of annual net sales thresholds. We have also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to KPL-716. Additionally, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

Under our license agreement with Novo Nordisk, we are obligated to make future milestone payments upon the achievement of specified clinical, regulatory, initial sales milestones as well as upon the achievement of annual net sales thresholds, including a payment of $1.0 million upon the earlier to occur of a specified regulatory milestone and January 2020. We are also obligated to pay royalties on annual net sales of products licensed under the agreement. In addition, we are obligated to make a payment upon the completion of technology transfer.

We have an exclusive option to acquire all outstanding capital stock of Primatope, which, subject to extension and the payment of specified extension fees, is exercisable until January 2019. If the option is exercised, we will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10.0 million as well as potential milestone payments of up to $10.0 million, in each case payable in a combination of cash and issuance of our Class A common shares.

Critical Accounting Policies and Significant Judgments and Estimates

Our consolidated financial statements are prepared in accordance with generally accepted accounting principles in the United States. The preparation of our consolidated financial statements and related disclosures requires us to make estimates and judgments that affect the reported amounts of assets, liabilities, costs and expenses, and the disclosure of contingent assets and liabilities in our financial statements. We base our estimates on historical experience, known trends and events and various other factors that we believe are reasonable under the circumstances, the results of which form the basis for making judgments about the carrying values of assets and liabilities that are not readily apparent from other sources. We evaluate our estimates and assumptions on an ongoing basis. Our actual results may differ from these estimates under different assumptions or conditions.

While our significant accounting policies are described in more detail in Note 2 to our consolidated financial statements appearing at the end of this prospectus, we believe that the following accounting policies are those most critical to the judgments and estimates used in the preparation of our consolidated financial statements.

Accrued Research and Development Expenses

As part of the process of preparing our consolidated financial statements, we are required to estimate our accrued research and development expenses. This process involves reviewing open contracts and purchase orders, communicating with our personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of actual costs. The majority of our service providers invoice us in arrears for services performed, on a pre-determined schedule or when contractual milestones are met; however, some require advanced payments. We make estimates of our accrued expenses as of each balance sheet date in the consolidated financial statements based on facts and circumstances known to us at that time. We periodically confirm the accuracy of these estimates with the service providers and make adjustments if necessary. Examples of estimated accrued research and development expenses include fees paid to:

- vendors, including research laboratories, in connection with pre-clinical development activities;
- CROs and investigative sites in connection with pre-clinical studies and clinical trials; and
- CMOs in connection with drug substance and drug product formulation of pre-clinical studies and clinical trial materials.

We base our expenses related to pre-clinical studies and clinical trials on our estimates of the services received and efforts expended pursuant to quotes and contracts with multiple research institutions and CROs that conduct and manage pre-clinical studies and clinical trials on our behalf. The financial terms of these agreements are subject to negotiation, vary from contract to contract and may result in uneven payment flows. There may be instances in which payments made to our vendors will exceed the level of services provided and result in a prepayment of the expense. Payments under some of these contracts depend on factors such as the successful enrollment of patients and the completion of clinical trial milestones. In accruing service fees, we estimate the time period over which services will be performed and the level of effort to be expended in each period. If the actual timing of the performance of services or the level of effort varies from the estimate, we adjust the accrual or the amount of prepaid expenses accordingly. Although we do not expect our estimates to be materially different from amounts actually incurred, our understanding of the status and timing of services performed relative to the actual status and timing of services performed may vary and may result in reporting amounts that are too high or too low in any particular period. To date, there have not been any material adjustments to our prior estimates of accrued research and development expenses.

Share-Based Compensation

We measure options and other share-based awards granted to employees and directors based on the fair value on the date of the grant and recognize the corresponding compensation expense of those awards over the requisite service period, which is generally the vesting period of the respective award. We have only issued options and restricted share awards with service-based vesting conditions and record the expense for these awards using the straight-line method.

For share-based awards granted to consultants and non-employees, we recognize compensation expense over the period during which services are rendered by such consultants and non-employees until completed. At the end of each financial reporting period prior to completion of the service, the fair value of these awards is remeasured using the then-current fair value of our Class A common shares and updated assumption inputs in the Black-Scholes option-pricing model.

We estimate the fair value of each option grant using the Black-Scholes option-pricing model, which uses as inputs the fair value of our Class A common shares and assumptions we make for the volatility of our Class A common shares, the expected term of our options, the risk-free interest rate for a period that approximates the expected term of our options and our expected dividend yield.

Determination of the Fair Value of Common Shares

As there has been no public market for our Class A common shares to date, the estimated fair value of our common shares has been determined by our board of directors as of the date of each option grant, with input from management, considering third-party valuations of our Class A common shares as well as our board of directors' assessment of additional objective and subjective factors that it believed were relevant and which may have changed from the date of the most recent third-party valuation through the date of the grant. These third-party valuations were performed in accordance with the guidance outlined in the American Institute of Certified Public Accountants' Accounting and Valuation Guide, *Valuation of Privately-Held-Company Equity Securities Issued as Compensation*. Our common share valuations were prepared using the option-pricing method, or OPM, which used a market approach to estimate our enterprise value. The OPM treats common shares and preferred shares as call options on the total equity value of a company, with exercise prices based on the value thresholds at which the allocation among the various holders of a company's securities changes. Under this method, the common shares have value only if the funds available for distribution to shareholders exceeded the value of the preferred share liquidation preferences at the time of the liquidity event, such as a strategic sale or a merger. A discount for lack of marketability of the common shares is then applied to arrive at an indication of value for the common shares. These third-party valuations were performed at various dates, which resulted in valuations of our common shares of $1.59 per share as of October 15, 2015, $1.86 per share as of September 16, 2016, $3.80 per share as of March 8, 2017 and June 29, 2017, $4.46 per share as of September 4, 2017, $4.68 as of December 14, 2017 and $10.36 as of March 1, 2018.

Our board of directors considered various objective and subjective factors to determine the fair value of our Class A common shares as of each grant date, including:

- the prices at which we sold preferred shares and the superior rights and preferences of the preferred shares relative to our Class A common shares at the time of each grant;
- the progress and value of our research and development programs, including the status of pre-clinical studies and planned clinical trials for our product candidates;
- our stage of development and our business strategy;
- external market conditions affecting the biotechnology industry, and trends within the biotechnology industry;
- our financial position, including cash on hand, our historical and forecasted performance and operating results;
- the lack of an active public market for our Class A common shares and our preferred shares;
- the likelihood of achieving a liquidity event, such as an initial public offering, or IPO, or a sale of our company in light of prevailing market conditions;
- the analysis of IPOs and the market performance of similar companies in the biopharmaceutical industry;
- the market value of equity interests in similar corporations and other entities engaged in businesses substantially similar to ours;

- the most recent transactions involving our preferred shares;
- current and potential strategic relationships, licenses and acquisitions; and
- competitive developments.

The assumptions underlying these valuations represent management's best estimates, which involve inherent uncertainties and the application of management judgment. As a result, if factors or expected outcomes change and we use significantly different assumptions or estimates, our share-based compensation expense could be materially different.

Following the closing of this offering, the fair value of our Class A common shares will be determined based on the quoted market price of our Class A common shares.

Options Granted

The following table sets forth by grant date the number of Class A common shares subject to options granted from January 1, 2016 through the date of this prospectus, the per share exercise price of the options, the fair value of common shares per share on each grant date and the per share estimated fair value of the options:

Grant Date	Number of Shares Subject to Options Granted	Per Share Exercise Price of Options	Fair Value per Common Share on Grant Date	Per Share Estimated Fair Value of Options
March 16, 2016	27,445	$ 1.59	$ 1.59	$ 1.05
June 1, 2016	22,322	$ 1.59	$ 1.59	$ 0.96
September 14, 2016	246,420	$ 1.86	$ 1.86	$ 1.18
December 7, 2016	33,486	$ 1.86	$ 1.86	$ 1.22
March 6, 2017	39,341	$ 3.80	$ 3.80	$ 2.54
June 7, 2017	42,815	$ 3.80	$ 3.80	$ 2.50
June 29, 2017	1,275,144	$ 3.80	$ 3.80	$ 2.51
September 14, 2017	78,685	$ 4.46	$ 4.46	$ 2.92
December 14, 2017	110,889	$ 4.68	$ 4.68	$ 3.07
March 1, 2018	1,652,321	$ 10.36	$ 10.36	$ 7.17

Emerging Growth Company Status

The JOBS Act permits an "emerging growth company" such as us to take advantage of an extended transition period to comply with new or revised accounting standards applicable to public companies until those standards would otherwise apply to private companies. We have irrevocably elected to "opt out" of this provision and, as a result, we will comply with new or revised accounting standards when they are required to be adopted by public companies that are not emerging growth companies.

Off-Balance Sheet Arrangements

We did not have during the periods presented, and we do not currently have, any off-balance sheet arrangements, as defined in the rules and regulations of the Securities and Exchange Commission.

Recently Issued Accounting Pronouncements

A description of recently issued accounting pronouncements that may potentially impact our financial position and results of operations is disclosed in Note 2 to our consolidated financial statements appearing at the end of this prospectus.

Quantitative and Qualitative Disclosures about Market Risks

Interest Rate Risk

As of December 31, 2016, December 31, 2017 and March 31, 2018, we had cash equivalents consisting of money market funds and U.S. Treasury securities. Based on the carrying value of the cash equivalents, an immediate change by 100 basis points in the interest rates would not have a material impact on our financial position or results of operations.

BUSINESS

Overview

We are a clinical-stage biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have a pipeline of product candidates across various stages of development, currently focused on autoinflammatory and autoimmune conditions. We have three clinical-stage product candidates, one of which is anticipated to commence a Phase 3 clinical trial in 2018. We follow a disciplined and methodical approach to selectively identify and acquire product candidates with strong biologic rationales or validated mechanisms of action. We believe that each of our product candidates has the potential to address multiple indications.

Our portfolio of product candidates offers multiple development opportunities. By modulating different parts of the innate and adaptive immune system, these product candidates together have the potential to provide a variety of mechanisms to address multiple devastating diseases.

- **Rilonacept** (ARCALYST) is a protein cytokine trap for inhibiting interleukin-1a, or IL-1a, and interleukin-1b, or IL-1b. Cytokines are small proteins that play a key role in cell signaling. Rilonacept is approved by the U.S. Food and Drug Administration, or FDA, for the treatment of cryopyrin-associated periodic syndromes, or CAPS, and has been commercially available from Regeneron Pharmaceuticals, Inc., or Regeneron, for this indication since 2008. We are initially developing rilonacept for the treatment of recurrent pericarditis, a debilitating inflammatory cardiovascular disease. We are not aware of any therapy currently approved by the FDA for the treatment of recurrent pericarditis. We are currently conducting an open-label Phase 2 proof-of-concept clinical trial in this disease and expect to report preliminary data in 2018. The trial is divided into five parts to evaluate rilonacept in various subgroups of patients with pericarditis. As of April 23, 2018, a total of four subjects were enrolled in the clinical trial. Three of the four subjects were experiencing symptomatic recurrent pericarditis at the time of enrollment. All three subjects showed a reduction in C-reactive protein measurements, as well as a reduction in scores of an 11-point Numerical Rating Scale instrument for assessing pericardial-associated chest pain, in the first week of treatment. The fourth subject, in a separate patient cohort, was on corticosteroids at the time of enrollment but not experiencing active symptoms, and had a history of corticosteroid dependence. This subject was treated with rilonacept and as of April 23, 2018, had been free of pericarditis flares for three weeks. Among these subjects, the most commonly reported adverse reaction associated with rilonacept has been injection-site reactions.

- **Mavrilimumab** is a monoclonal antibody that antagonizes the signaling of granulocyte macrophage colony stimulating factor, or GM-CSF. We are focusing our initial development efforts for mavrilimumab on giant cell arteritis, or GCA, an inflammatory disease of the blood vessels with unmet medical need that can lead to blindness if left untreated. MedImmune Limited, or MedImmune, initially developed mavrilimumab for the treatment of rheumatoid arthritis, or RA. MedImmune's Investigational New Drug application, or IND, for the clinical development of mavrilimumab for the treatment of RA was initially put on clinical hold in 2010 before human data had been generated due to certain effects that were observed in non-clinical studies, which coincides with a theoretical risk of developing pulmonary alveolar proteinosis, or PAP, possibly in the setting of GM-CSF inhibition. Since then, in 2014, the FDA acknowledged that clinical studies in refractory RA may be appropriate based on MedImmune's clinical studies in Europe in which it dosed over 550 RA patients with mavrilimumab with no evidence of PAP. MedImmune has since withdrawn the IND for mavrilimumab for the treatment of RA. We intend to develop mavrilimumab for the treatment

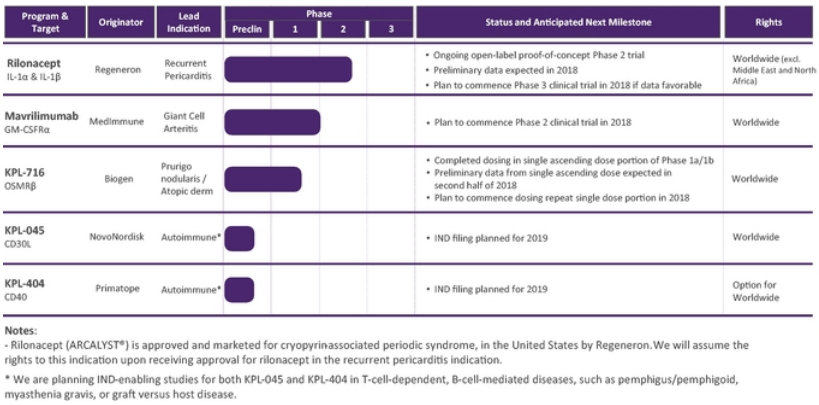
of GCA under a new IND in the United States and new Clinical Trial Application, or CTA, in Europe, and plan to initiate a Phase 2 clinical trial in 2018.

- **KPL-716** is a monoclonal antibody that simultaneously inhibits the signaling of the cytokines interleukin-31, or IL-31, and oncostatin M, or OSM, by targeting their common receptor subunit, oncostatin M receptor beta, or OSMRb. We plan to study KPL-716 in a variety of pruritic and fibrotic indications driven by these cytokines and we believe KPL-716 is the only monoclonal antibody in development that simultaneously targets both pathways. We are currently enrolling subjects in a Phase 1a/1b clinical trial in healthy volunteers and in subjects with atopic dermatitis as a proof-of-concept for pruritic conditions. We have completed dosing in the first portion of the Phase 1a/1b clinical trial utilizing a single ascending dose design. Each dose cohort in the first portion received a single dose of KPL-716 administered either intravenously or subcutaneously. We expect to report preliminary data from the single ascending dose cohorts of this portion of the trial in the second half of 2018. In the second portion of the Phase 1a/1b clinical trial, each subject in the cohort will receive repeated single doses of KPL-716 administered subcutaneously. We plan to commence dosing in this portion of the trial in 2018, subject to submission to, and review by, the FDA and Canadian regulatory authorities of our recently completed primate chronic toxicology study. If the data from the Phase 1a/1b clinical trial are favorable, we expect our two initial targeted indications for future development of KPL-716 to be prurigo nodularis and atopic dermatitis, both inflammatory, pruritic skin conditions with unmet medical need.

- **KPL-045** is a monoclonal antibody inhibitor of the CD30/CD30L interaction, a T-cell co-stimulatory receptor involved in activated T-memory cell function. We are planning IND-enabling studies in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in 2019.

- **KPL-404** is a monoclonal antibody inhibitor of the CD40/CD40L interaction, a central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity. We are planning IND-enabling studies in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in 2019.

The following table summarizes our current pipeline of product candidates:



Program & Target	Originator	Lead Indication	Phase	Status and Anticipated Next Milestone	Rights
Rilonacept IL-1α & IL-1β	Regeneron	Recurrent Pericarditis	Preclin / 1 / 2 / 3	• Ongoing open-label proof-of-concept Phase 2 trial • Preliminary data expected in 2018 • Plan to commence Phase 3 clinical trial in 2018 if data favorable	Worldwide (excl. Middle East and North Africa)
Mavrilimumab GM-CSFRα	MedImmune	Giant Cell Arteritis	Preclin / 1 / 2 / 3	• Plan to commence Phase 2 clinical trial in 2018	Worldwide
KPL-716 OSMRβ	Biogen	Prurigo nodularis / Atopic derm	Preclin / 1 / 2 / 3	• Completed dosing in single ascending dose portion of Phase 1a/1b • Preliminary data from single ascending dose expected in second half of 2018 • Plan to commence dosing repeat single dose portion in 2018	Worldwide
KPL-045 CD30L	NovoNordisk	Autoimmune*	Preclin / 1 / 2 / 3	• IND filing planned for 2019	Worldwide
KPL-404 CD40	Primatope	Autoimmune*	Preclin / 1 / 2 / 3	• IND filing planned for 2019	Option for Worldwide

Notes:
- Rilonacept (ARCALYST®) is approved and marketed for cryopyrinassociated periodic syndrome, in the United States by Regeneron.We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication.

* We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.

In addition to the indications described above, we plan to evaluate rilonacept, mavrilimumab and KPL-716 in other indications. We plan to be opportunistic in our business development activities to identify and potentially acquire the rights to additional programs that expand our existing portfolio. We have also initiated our own internal research efforts to discover and develop molecules to address areas of unmet medical need.

We intend to directly commercialize our product candidates, if approved, in the United States and select international markets. In parallel with our product development timelines, we plan to build our own commercial and operational organizations around the world. We anticipate building targeted medical affairs and sales teams focused on specialist physicians who treat the patient populations addressed by our product candidates.

Our Team

We have assembled an experienced management team with a successful track record, many of whom have previously worked together. Our team has expertise across the spectrum of global drug discovery, development, manufacturing and commercialization activities in diseases within both large and orphan indications. Our Chairman and Chief Executive Officer, Sanj K. Patel, has more than 25 years of scientific, clinical and commercial experience in the pharmaceutical and biotechnology industries. Our Chief Medical Officer, John F. Paolini, M.D., Ph.D., has more than 15 years of experience planning, operating and executing clinical development programs across a range of disease indications from orphan diseases to large cardiovascular diseases, and ten years as a practicing cardiologist. Other members of our senior management team have held key management positions at other companies that developed and commercialized therapies for underserved, rare and specialty-focused patient populations. These companies include Synageva, Genzyme, Novo Nordisk, Shire, Sanofi, Pfizer, Bayer, Merck, Novartis and Vertex, among others.

Our Strategy

Our vision is to build a fully-integrated global biopharmaceutical company by discovering, acquiring, developing and commercializing life-changing therapies for debilitating diseases. We are currently developing a pipeline of novel drug product candidates for the treatment of

autoinflammatory and autoimmune diseases, and we aim to be an industry leader in these areas. We are pursuing multiple programs in parallel, with the goal of delivering safe and effective therapies to patients as efficiently as possible.

Critical components of our business strategy include the following:

- **Efficiently and rapidly advance our product candidates through the development process.** We believe that our product candidates have the potential to address significant unmet medical needs and intend to develop them as efficiently and rapidly as possible. In 2018, we expect to report preliminary Phase 2 data for rilonacept and, if the Phase 2 data are favorable, we plan to initiate a Phase 3 clinical trial for rilonacept in recurrent pericarditis. For KPL-716, we anticipate reporting preliminary data from the single dose cohorts in our Phase 1a/1b clinical trial in normal healthy volunteers and subjects with atopic dermatitis in the second half of 2018. We also expect to initiate a Phase 2 clinical trial of mavrilimumab in GCA in 2018.

- **Commercialize our product candidates to bring new or improved therapies to patients in need.** We intend to market and commercialize our product candidates, if approved, in the United States and select international markets by developing our own sales, marketing, medical affairs and reimbursement organizations. We anticipate creating a targeted sales organization that supports specialist physicians who treat these specific patient populations and plan to build out this organization as our product candidates approach potential regulatory approval. We believe this approach will allow us to effectively reach patients and prescribers that our product candidates target and leverage the commercial potential of our product candidates.

- **Maximize our existing portfolio opportunity by expanding use across multiple indications.** A core component of our approach to product development is identifying assets that each have the potential to treat multiple diseases. We aim to develop and commercialize our product candidates to produce meaningful impact for patients across all relevant indications. Our assets are designed to modulate signaling pathways that are implicated across a spectrum of autoimmune and autoinflammatory conditions. For example, our lead product candidate, rilonacept, is being studied in recurrent pericarditis, and we believe it may be effective in other IL-1a-mediated diseases characterized by painful serosal inflammation. We also believe that both mavrilimumab and KPL-716 have potential in additional indications.

- **Leverage our value-driven approach to identify, acquire, discover and develop new therapies.** We follow a disciplined and methodical approach to our review of new opportunities. We focus on research-based and comprehensive indication mapping exercises to categorize and prioritize indications of interest. We evaluate a variety of factors for potential product candidates and discovery targets, including biologic rationale for addressing the disease, potential for regulatory approval, commercial viability, intellectual property position, prospects for favorable pricing and reimbursement and the impact of competition. We also look at assets that could potentially address multiple indications. In building our current pipeline, we evaluated a large number of opportunities and negotiated agreements with parties for the assets that met our criteria and have acquired the rights to develop and commercialize five separate biologics. Going forward, we intend to be opportunistic in our business development activities.

- **Build our core capability in autoimmune and autoinflammatory diseases to establish a leadership position in the field.** Our current pipeline consists of protein therapeutics across various stages of drug development, including a cytokine trap, rilonacept, and four monoclonal antibodies—mavrilimumab, KPL-716, KPL-045 and KPL-404. Both categories of

therapeutics functionally inhibit signaling pathways that are implicated in autoinflammatory- or autoimmune-driven pathologies. We intend to leverage our internal discovery efforts and business development capabilities to complement our existing portfolio to build our core capability and establish a leadership position in the field.

Our Product Candidates

Rilonacept (ARCALYST)

Overview

Rilonacept was approved by the FDA for the treatment of cryopyrin-associated periodic syndrome, or CAPS, which includes cold auto-inflammatory syndrome and Muckle-Wells syndrome, and has been commercially available in the United States by Regeneron for this indication since 2008. We licensed rilonacept in 2017 from Regeneron. We believe that rilonacept has potential to treat certain diseases mediated by both IL-1a and IL-1b. Our lead indication for rilonacept is recurrent pericarditis, which is a recurring painful inflammation of the pericardium. We have initiated an open-label Phase 2 proof-of-concept clinical trial in patients with recurrent pericarditis who are not well controlled by, or who cannot be weaned off of, current standard of care. We expect to report preliminary data from this trial in 2018. If the results from this trial are favorable, we plan to initiate a Phase 3 clinical trial in the United States for rilonacept in 2018. We also plan to evaluate rilonacept in additional diseases mediated by IL-1a.

There is currently one other FDA-approved agent that blocks both IL-1a and IL-1b signaling, anakinra, and one that blocks only IL-1b, canakinumab. We believe both therapies have limitations, and neither is approved by the FDA for treatment of pericarditis. Anakinra requires once-daily injections, and canakinumab only blocks IL-1b, making it less effective or ineffective in diseases driven by IL-1a pathology. We believe that rilonacept with its more moderate, once-weekly dosing schedule and its ability to inhibit both IL-1a and IL-1b could provide an improved therapeutic option for a variety of IL-1a-mediated diseases.

Mechanism of Action

Rilonacept is an inhibitor of IL-1a and IL-1b. IL-1a and IL-1b have been demonstrated to play a key role in inflammatory diseases. IL-1a and IL-1b provoke potent, pro-inflammatory events by engaging the IL-1a and IL-1b receptor. Following tissue insult, the release of IL-1a acts as the primary initiating signal to coordinate the mobilization of immune cells to the damaged area, while IL-1b is secreted mostly by macrophages and is a prototypical cytokine of the canonical inflammasome. IL-1a and IL-1b signaling results in a dramatic increase in the production of cytokines that orchestrate the proliferation and recruitment of phagocytes to the site of damage, resulting in inflammation. Moreover, IL-1a and IL-1b signaling also affect other immune-system cells, such as T-cells and B-cells.

IL-1b's role in the inflammation process has been extensively studied, while in comparison, much is still unknown about the independent function of IL-1a in disease pathology. Despite driving similar immunological outcomes, IL-1a and IL-1b differ substantially in their expression and regulation, and non-redundant roles for IL-1a or IL-1b have been demonstrated in multiple inflammatory diseases. There are disease states in which IL-1b inhibition alone does not appear to be sufficient for disease remission in the absence of IL-1a inhibition. Published studies suggest certain autoinflammatory diseases may, in fact, be pathologically driven primarily by IL-1a.

An investigator-initiated study of anakinra successfully demonstrated mechanistic proof-of-concept for inhibiting both IL-1a and IL-1b in the treatment of recurrent pericarditis. In a published case study, a patient with a refractory form of recurrent pericarditis, who was

well-controlled on anakinra, was switched from anakinra to canakinumab, which inhibits only IL-1b, for tolerability reasons. The patient's disease returned despite further dose escalation of canakinumab. When the patient was switched back to anakinra, which inhibits IL-a and IL-b, the disease promptly went back into remission. These data, along with confirmatory market research, may indicate that IL-1a and IL-1b play unique roles in recurrent pericarditis and other autoinflammatory diseases in which the pathology may be driven primarily by IL-1a.

Background and Market Opportunity for Recurrent Pericarditis

Pericarditis is the most common disorder involving the pericardium, the two-layered sac that surrounds the heart. Pericarditis is an inflammation of this sac and is typically characterized by significant chest pain, shortness of breath, coughing and fatigue and is often misconstrued by patients as a heart attack. In addition, typical signs of pericarditis include pericardial friction rub, electrocardiogram changes or pericardial effusion, which is a build-up of fluid around the heart. Pericarditis is described as recurrent if, following an initial occurrence of pericarditis, it recurs after a symptom-free period of about four to six weeks. Pericarditis is considered chronic if symptoms of any one episode last longer than three months, typically causing significant pain and frustration. If pericarditis is left untreated, patients can develop thickening and scarring of the pericardium, potentially requiring invasive surgical stripping. Pericardial effusion, if large enough, can compress the heart externally, requiring emergent drainage.

We intend to focus our development of rilonacept for the treatment of recurrent pericarditis initially in the United States. Based on claims data from 2012 and literature sources, we estimate that there are approximately 90,000 newly incident patients with episodes of pericarditis in the United States per year. Approximately 20% to 30% of these patients experienced additional recurrent flares, and required additional treatment with non-steroidal anti-inflammatory drugs, or NSAIDs, the immunosuppressive drug colchicine or steroids, either alone or in combination. This results in an annual incidence of approximately 20,000 patients with recurrent pericarditis. Based on an average three year course of recurrent disease, we estimate there may be up to 60,000 patients with recurrent pericarditis in the United States and 5% of these recurrent patients are refractory to their currently available pharmacotherapy (approximately 3,000 patients in the United States). Additionally, we estimate that roughly an additional 9,000 recurrent patients are considered by their physicians not well-controlled on their current therapy, meaning that they are unable to wean off existing therapy, have multiple recurrences despite repeated pharmacotherapy or cannot tolerate their existing therapy.

There may be other thoracic inflammatory syndromes where rilonacept may prove beneficial, such as pericarditis associated with postpericardiotomy syndrome, an inflammatory reaction of the pericardium in patients who have undergone surgery that involves opening the pericardium. Postpericardiotomy syndrome occurs in up to 30% of the 300,000 patients in the United States undergoing open heart surgery, and we believe rilonacept may be a therapeutic option for a subset of these patients.

Current Treatment Landscape for Recurrent Pericarditis

We are not aware of any current therapies approved by the FDA for the treatment of recurrent pericarditis. A patient's initial acute episode of pericarditis is typically treated with over-the-counter or prescription NSAIDs or colchicine, both of which are used off-label. Recurrent episodes are treated in a similar manner or by adding systemic corticosteroids which are also used off-label. Both colchicine and corticosteroids often have deleterious effects when used at high doses or for long periods of time, including, for colchicine, gastrointestinal distress and neutropenia and, for corticosteroids, glaucoma, fluid retention, hypertension, mood changes, memory changes, other

psychological effects, weight gain and diabetes. Fourth-line treatment for these patients may include other immunosuppressants such as methotrexate and azothiaprine, as well as anakinra.

Our Solution

Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1a and IL-1b signaling. Beyond recurrent pericarditis, we believe there is significant potential for rilonacept to address additional indications, including other pericarditis populations. More broadly, we believe diseases characterized by painful serosal inflammation may be driven by IL-1a, and we intend to consider development of rilonacept in these indications and in others where we believe IL-1a or IL-1b play a key role in disease pathophysiology.

Clinical Development Plan for Recurrent Pericarditis

We have initiated an open-label Phase 2 proof-of-concept clinical trial to explore clinical and biochemical endpoints of pericarditis symptomatology and to collect inter- and intra-subject variability data on both at-baseline and on-treatment parameters. The trial is divided into five parts, each enrolling up to 10 subjects who are currently on any combination of co-administered NSAIDs, colchicine or corticosteroids. The subjects are dosed using the approved rilonacept dose for CAPS, which is a loading dose of two 160 mg subcutaneous doses (320 mg total), followed by single, self-administered 160 mg subcutaneous doses every seven days for a total of six weeks. This is followed by an 18-week extension period. During the extension period, the investigator may choose to wean concomitant NSAIDs, colchicine or corticosteroids according to standard-of-care paradigms.

The five parts of the trial with different patient populations are:

- Part 1: Symptomatic subjects with recurrent pericarditis receiving NSAIDS +/– colchicine +/– steroids with high CRP, a marker of inflammation, measurements;
- Part 2: Symptomatic subjects with recurrent pericarditis receiving NSAIDS +/– colchicine +/– steroids without elevated CRP measurements but with evidence of pericardial inflammation by MRI;
- Part 3: Subjects with recurrent pericarditis who are dependent upon or unable to wean off of corticosteroids;
- Part 4: Symptomatic subjects with postpericardiotomy syndrome receiving NSAIDS +/– colchicine +/– steroids with high CRP measurements; and
- Part 5: Subjects with postpericardiotomy syndrome who are dependent upon or unable to wean off of corticosteroids.

We intend to use the information gathered in this trial to confirm the results seen to date with anakinra and to inform an end-of-Phase 2 meeting with the FDA. The primary endpoint for Parts 1, 2 and 4 of the trial is to collect inter- and intra-subject variability data on CRP measurements and the NRS in subjects with symptomatic recurrent idiopathic pericarditis both at baseline and on treatment with rilonacept in order to inform power calculations for future trials in pericarditis. The primary endpoint for Parts 3 and 5 is to evaluate feasibility of weaning patients from corticosteroids while receiving rilonacept. This trial has been designed with the potential to examine the response of patients with pericarditis caused by a variety of underlying etiologies.

As of April 23, 2018, a total of four subjects have been enrolled in the clinical trial. Three of the four subjects have been enrolled in Part 1, and were experiencing symptomatic recurrent pericarditis at the time of enrollment. All three subjects showed a reduction in CRP measurements, as well as a reduction in NRS, in the first week of treatment, which was sustained as of April 23,

2018. The fourth subject, in a separate patient cohort, was on corticosteroids at the time of enrollment in Part 3 of the study but not experiencing active symptoms, and had a history of corticosteroid dependence. This subject was treated with rilonacept and as of April 23, 2018, had been free of pericarditis flares for three weeks. Among these subjects, the most commonly reported adverse reaction associated with rilonacept has been injection-site reactions. We have not enrolled any subjects in Parts 2, 4 or 5 as of April 23, 2018.

Clinical History of Rilonacept

Regeneron evaluated rilonacept in a total of 21 clinical trials, including two trials in over 100 patients for the treatment of CAPS, and six trials in over 1,800 patients for the treatment of gout flares.

- CAPS: Regeneron evaluated rilonacept for the treatment of CAPS in two trials. In these trials, 109 patients with CAPS, including eight pediatric patients, were treated with at least one dose of rilonacept. In the pivotal efficacy trial, which evaluated the long-term efficacy and safety of once-weekly dosing, 160 mg of rilonacept markedly decreased the clinical signs and symptoms of CAPS.

- Gout: Regeneron evaluated rilonacept for the treatment of gout flares in six trials. In the two pivotal efficacy trials in patients with gout, which evaluated the efficacy of once-weekly dosing for the prevention of gout flares during initiation of uric acid-lowering therapy, rilonacept at doses of 80 mg and 160 mg significantly decreased the number of gout flares. Regeneron abandoned active development for the treatment of gout flares after receiving a complete response letter from the FDA requesting additional clinical data, as well as additional CMC information related to a proposed new dosage form Regeneron was evaluating for gout, which was different than the dosage form approved in the CAPS indication and now being used for pericarditis.

- Other Indications: Regeneron conducted a total of 13 clinical trials of rilonacept for the treatment of rheumatoid arthritis, or RA, polymyalgia rheumatica, osteoarthritis, coronary artery disease, systemic juvenile idiopathic arthritis and end-stage renal disease.

In the 21 clinical studies conducted by Regeneron with rilonacept to date, the most common adverse events reported were injection site reactions and upper respiratory tract infections. Across these studies, there were a total of five serious adverse events, or SAEs, that were assessed by investigators as drug related. Among patients treated with rilonacept there were three SAEs, colitis, gastrointestinal haemorrhage, and drug eruption. One patient treated with placebo experienced cellulitis and another placebo-treated patient died. The largest clinical programs conducted by Regeneron with rilonacept were its Phase 2 and Phase 3 programs for gout flare prevention, which treated a total of 1,886 patients. The most common adverse events reported for the 160 mg dose, the dosage used for the treatment of CAPS, were injection site reactions (15.5% for rilonacept versus 2.6% for placebo) and upper respiratory tract infections (10.3% for rilonacept versus 10.1% for placebo).

Mavrilimumab

Overview

Mavrilimumab is a fully-human monoclonal antibody that antagonizes GM-CSF signaling by binding to the alpha subunit of the GM-CSF receptor. Our lead indication for mavrilimumab is GCA, an inflammatory disease of blood vessels, for which we plan to initiate a Phase 2 trial in 2018. We also plan to evaluate mavrilimumab in additional diseases for patients with high unmet medical need, where we believe there is a strong mechanistic rationale.

Before we licensed mavrilimumab in 2017, MedImmune, Limited, or MedImmune, was developing mavrilimumab for the treatment of RA.

Mechanism of Action

Mavrilimumab is designed to inhibit the signaling of GM-CSF, a growth factor that stimulates the production of certain types of white blood cells. Studies have demonstrated that with GM-CSF overexpression, pathological changes almost always follow. Reported data suggest GM-CSF is a key player in autoinflammation and autoimmunity, as follows:

- GM-CSF enhanced trafficking of myeloid cells through activated endothelium of blood vessels and contributed to monocyte and macrophage accumulation in blood vessels during inflammation;
- GM-CSF promoted activation, differentiation, survival and proliferation of monocytes and macrophages, as well as resident tissue macrophages in inflamed tissues;
- GM-CSF production led to activation of the vasculature and bone marrow and also promoted the differentiation of effector T cells at inflamed sites and draining lymph nodes; and
- GM-CSF regulated the phenotype of antigen-presenting cells in inflamed tissues by promoting the differentiation of infiltrating monocytes into M1 macrophages and monocyte-derived dendritic cells, or MoDCs.

Additionally, GM-CSF has been shown to be a confirmed mediator in RA based on the results from the Phase 2b clinical trial in RA conducted by MedImmune. In this trial, mavrilimumab achieved the co-primary endpoints of change from baseline in disease activity score, or DAS, at week 12 and a response of 20% or greater improvement in the American College of Rheumatology criteria, at week 24. Patients with mavrilimumab showed a statistically significant reduction in DAS scores at all dosages compared to placebo, and significantly more mavrilimumab-treated patients achieved ACR20 at all dosages compared to placebo.

Background and Market Opportunity for Giant Cell Arteritis

GCA is an inflammatory disease of the blood vessels that strikes older adults and causes headaches, jaw and other muscle claudication, and possible ischemic visual loss. Many of the symptoms and signs of GCA result from involvement of the cranial branches of arteries that originate from the aortic arch, but the disease is systemic, and vascular involvement can be widespread. GCA is characterized by infiltration of monocytes, macrophages and the formation of giant cells (i.e., multinucleated fusions of macrophages). GCA generally occurs in adults over 50 years old with a 3:1 imbalance of women to men. We estimate there to be approximately 75,000 to 150,000 prevalent patients with GCA in the United States with similar prevalence rates for other major markets and believe that the incidence of GCA will increase over time as the population ages.

Current Treatment Landscape for Giant Cell Arteritis

Glucocorticoids, a type of corticosteroid, are the mainstay for the treatment of GCA because they normalize inflammatory markers and resolve patient symptoms. Many patients receive long courses of this therapy to prevent disease flare-up, which are associated with significant and serious side effects, including glaucoma, fluid retention, hypertension, mood changes, memory changes, other psychological effects, weight gain and diabetes. Up to 80% of patients suffer from glucocorticoid toxicity as a result of GCA treatment.

Despite being effective for some patients, many are unable to wean off of corticosteroids because they continue to experience disease flares as the dose is reduced. In one study cohort published in the literature that followed 106 patients with GCA for 4.5 to 10.1 years, 68 patients (64%) experienced at least one relapse during or after weaning, and 38 patients (36%) experienced

two or more. Experimental evidence in mice suggests that corticosteroid treatment does not adequately suppress tissue-infiltrating macrophage function, a key cell type generated and maintained by GM-CSF signaling, and may explain why many patients require long-term chronic treatment and are unable to wean off corticosteroids. We believe by blocking GM-CSF signaling, mavrilimumab may provide additional benefit to these patients by reducing long-term sequelae that results from chronic vessel inflammation.

In addition, tocilizumab, an inhibitor of interleukin-6, or IL-6, is approved in the United States in GCA for use on top of a concomitant corticosteroid taper. However, nearly half of the patients studied in the Phase 3 clinical trial for tocilizumab experienced disease flares during the 52 weeks treatment period that included a 26-week corticosteroid taper. We believe this indicates a persistent unmet medical need.

Our Solution

We chose GCA as our first indication for mavrilimumab due to the mechanistic rationale of inhibiting GM-CSF. GM-CSF is a key growth factor for many of these key inflammatory cell types and is found in high concentrations at the site of damage in the vessel wall. We believe these data provide a solid rationale for antagonizing this signaling with mavrilimumab.

Phase 2 Clinical Trial for GCA

We plan to initiate a Phase 2 clinical trial of mavrilimumab for the treatment of GCA in 2018 in Europe after we submit an investigational medicinal product dossier, or IMPD, and a clinical trial application, or CTA, to and receive clearance from the competent authorities in Europe. We also intend to file an IND for studying mavrilimumab to treat GCA in the United States, which will first need to be cleared by the FDA. Our current plans include enrolling 30 to 60 newly diagnosed and refractory GCA patients who will be randomized to mavrilimumab versus placebo on top of a corticosteroid taper. The primary endpoint will quantify maintenance of complete remission without clinical signs or symptoms of GCA in subjects treated with mavrilimumab versus placebo.

After initiating the planned Phase 2 trial in GCA, we anticipate initiating research and development activities of mavrilimumab's potential across other various disease states where cells of myeloid phenotype have been implicated by the literature, such as other vasculitides and cardiomyopathies, diseases characterized by barrier dysfunction, other arthropathies or oncologic indications.

Clinical History in Rheumatoid Arthritis

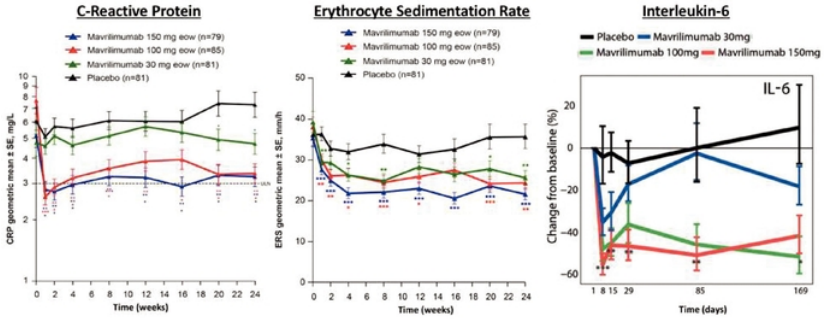
MedImmune had received authorization to conduct clinical trials for rheumatoid arthritis, or RA, in Europe and executed an extensive Phase 1 and Phase 2 clinical program where the company studied mavrilimumab in over 550 patients with RA through Phase 2b. All of these clinical trials achieved their prospectively defined primary endpoints of safety or efficacy.

In the United States, MedImmune did not finish its IND process with the FDA after the IND was initially put on clinical hold in 2010 before any human data had been generated. Certain effects had been observed in the lungs of non-human primates, which coincides with the theoretical risk of pulmonary alveolar proteinosis, or PAP, possibly developing in the setting of GM-CSF inhibition. Subsequently, MedImmune discussed its Phase 1 and initial Phase 2b clinical findings with the FDA, and the FDA acknowledged in November 2014 that a clinical trial of mavrilimumab in the United States may be appropriate in patient populations with high morbidity and limited treatment options, including refractory RA. MedImmune did not engage in further dialogue with the FDA and has since withdrawn the IND for mavrilimumab for the treatment of RA, and we plan to file a new IND for our planned Phase 2 clinical trial in GCA. MedImmune opted to out-license the program due to reasons related to its overall portfolio strategy and focus on its three main therapeutic areas,

which will allow us as the licensee to continue the dialogue directly with the FDA in the context of our proposed clinical development program. MedImmune's European clinical trials in RA, which included an extensive pulmonary safety adjudication program, revealed no signals of altered pulmonary function (including PAP) attributable to mavrilimumab following long-term administration. We believe that these long-term data, which have not yet been presented to the FDA, support that PAP is a theoretical risk and confirm our belief that mavrilimumab is appropriate to study in patients with diseases like GCA with high morbidity and limited treatment options. Also, several other molecules that inhibit the GM-CSF pathway (or inhibitors of its signaling pathway such as janus kinase 2), have commenced clinical studies in other indications in the United States since 2010 which may provide additional support for our discussions with FDA.

We believe that the trials conducted by MedImmune provide substantial support for the potential of mavrilimumab in autoimmune diseases. In these trials, mavrilimumab was observed to be well-tolerated. The most common adverse event was infection, with all dose groups (30 mg, 100 mg, 150 mg) in a Phase 2b clinical trial reporting similar rates of infection compared to the placebo group. We believe that these safety results provide an accurate early representation of the safety profile of mavrilimumab, which we believe to be at least competitive with and potentially better than existing systemically administered agents for autoimmune diseases.

Mavrilimumab's results from Phase 2b clinical trials in RA have provided important information about its safety and efficacy profile and helped solidify our choice for focusing our development efforts in GCA as a lead indication. In addition to the reductions to the primary endpoint demonstrated in the Phase 2b trials, other markers of inflammation, such as CRP, erythrocyte sedimentation rate, or ESR, and IL-6, were similarly reduced, as shown in the graphs below. CRP, ESR and IL-6 are key markers of disease activity for GCA. We believe that these results may also provide evidence for mavrilimumab's utility across a broad range of indications with a similar biomarker profile.



KPL-716 Overview

KPL-716 is a fully-human monoclonal antibody that targets OSMRb, which mediates signaling of IL-31 and OSM, two key cytokines implicated in inflammation, pruritus and fibrosis. We believe KPL-716 to be the only monoclonal antibody in development that targets both pathways simultaneously. We are initially developing KPL-716 for the treatment of prurigo nodularis and atopic dermatitis, both diseases where OSMRb signaling has been implicated. A significant portion of individuals in the United States experience at least one atopic pruritic disease during their lifetime, and it is well understood that most patients with one type of atopic condition tend to present with other allergic conditions. While we believe KPL-716 may be effective across many pruritic and

fibrotic diseases, we have prioritized our development efforts based on unmet medical need and potential market opportunity. We are currently conducting a Phase 1a/1b clinical trial of KPL-716 in healthy volunteers and in subjects with atopic dermatitis as a surrogate for a range of pruritic diseases. If the results of this Phase 1a/1b clinical trial are favorable, we plan to initiate further trials in prurigo nodularis and atopic dermatitis. We expect to report preliminary data from the single dose cohorts in our Phase 1a/1b clinical trial in the second half of 2018.

We acquired the assets relating to KPL-716 from Biogen MA, Inc., or Biogen, in 2016.

Mechanism of Action

The OSMRb subunit is an IL-6 type receptor which combines with one of two other subunits to form two distinct cytokine receptors used for the signaling of two different cytokines: IL-31, and OSM. IL-31 produced in the setting of an inflammatory response binds to the IL-31 receptor on keratinocytes, epidermal cells, leading to a sensation of pruritus and further inflammatory responses in the skin. In addition to interacting with IL-31 receptors on keratinocytes, IL-31 also stimulates pruritus directly through IL-31 receptors expressed on unmyelinated C-fibers in the skin responsible for the sensation and transmission of pruritic signaling.

OSM is produced primarily under inflammatory conditions and stimulates dermal fibroblast proliferation and migration as well as synthesis of collagen and glycosaminoglycan in the skin, leading to fibrosis. In addition to these functions, OSM signaling through the type II OSM receptor upregulates interleukin-4, or IL-4, interleukin-13 receptor, or IL-13Ra1, and interleukin-4 receptor, or IL-4Ra, in human skin equivalent cultures, upregulates IL-4Ra in primary human keratinocytes and also impairs expression of fillagrin, loricrin and involucrin (classical "differentiation" markers of the epidermal differentiation complex cluster) in human skin equivalent cultures. These data implicate OSM signaling as important in many autoimmune diseases characterized by barrier dysfunction, fibrosis and inflammation.

KPL-716 inhibits both IL-31 and OSM activities at their respective receptors, potentially disrupting the pruritus, inflammation and fibrosis mediated by these cytokine pathways.

Background and Market Opportunity for Prurigo Nodularis and Atopic Dermatitis

Prurigo Nodularis

Prurigo nodularis is a chronic inflammatory skin condition that affects primarily older adults and is characterized by multiple firm and extremely pruritic nodules typically located on the arms and legs. The etiology of prurigo nodularis is largely unknown, however, human biopsy studies have shown that IL-31, its receptor IL-31Ra, OSM and OSMRb are highly expressed in prurigo nodularis lesions. The pruritus is severe and distressing and can be sudden, sporadic or continuous, worsening with heat, sweating or irritation from clothing. The itching sensation in prurigo nodularis is extreme and often leads to scratching to the point of bleeding, infection or pain. Our market research to-date with physicians and patients highlights the severe and debilitating nature of this disease and the significant levels of unmet need. Multiple physicians have reported suicidal tendencies among their prurigo nodularis patients due to an overwhelming inability to control the unrelenting itch. The exact prevalence of prurigo nodularis is unknown, however, we estimate there to be approximately 300,000 prevalent cases in the United States.

Atopic Dermatitis

Atopic dermatitis is a chronic inflammatory skin disease that affects approximately 18 million adults in the United States. Human biopsy studies have shown that IL-31, its receptor IL-31Ra, OSM and OSMRb are highly expressed in atopic dermatitis lesions. Based upon public data analyses and discussions with physicians and key opinion leaders in the field, we estimate that

approximately 300,000 atopic dermatitis patients in the United States are diagnosed with a moderate-to-severe form of this disease that significantly impairs their professional and social life on a daily basis.

Current Treatment Landscape for Prurigo Nodularis and Atopic Dermatitis

Prurigo Nodularis

We are not aware of any current FDA-approved therapies for treating prurigo nodularis, and the treatment approach ranges from topical corticosteroids and occlusive steroid containing bandages for more mild patients to systemic corticosteroid, ultraviolet phototherapy and systemic therapies such as thalidomide, methotrexate and cyclosporine for those patients who fail initial treatments. Patients have reported using opioid pain medications to attempt to control the disease in its most severe form.

Atopic Dermatitis

Current therapies for atopic dermatitis are generally focused on the topical use of non-biologic small molecules, however, dupilumab (subcutaneously injected antibody directed to inhibiting signaling through IL-4Rα) has recently been approved by the FDA for the treatment of atopic dermatitis.

Our Solution

KPL-716 is a fully-human monoclonal antibody that targets two key pathways for the development of pruritus, inflammation and fibrosis through inhibition of OSMRb. Chronic pruritic diseases are often characterized by a complex interplay among pruritus, inflammation and fibrosis. The pathogenesis of chronic pruritic diseases involves interlocking positive feedback loops in which pruritus causes scratch, and scratch causes reactive inflammation through mechanical disruption of the skin architecture. The decline in skin barrier function and resulting bacterial colonization or infection ultimately increase extracellular matrix formation and collagen deposition, leading to fibrosis. Fibrosis then begets more pruritus through disruption and dysregulation of sensory nerve fiber expression.

Current therapies target only one or two aspects of this complex pathophysiology and are inevitably limited in their effectiveness. Targeting only one pathway may address a single aspect of the symptomatology, e.g., pruritus, but not the full spectrum of the pathophysiologic components of the disease. This point is particularly relevant since OSM is upregulated in many chronic inflammatory skin diseases and synergistically interacts with pruritic and inflammatory pathways. Of particular relevance is the central role of OSM in inflammation and barrier function and its autocrine effects on type II OSM receptor in IL-31-dependent epidermal proliferation and remodeling as well as inflammation.

There is a relatively large body of literature linking inflammatory pruritic and inflammatory diseases to both IL-31 and OSM via signaling though OSMRb. KPL-716 has been specifically designed to target both pathways simultaneously and thus KPL-716 may disrupt this pathologic cycle in patients afflicted by prurigo nodularis and atopic dermatitis.

Pre-clinical Development

In our pre-clinical development program we have observed favorable pharmacokinetics and toxicology characteristics to support clinical development of KPL-716. KPL-716 has shown signs of efficacy in two non-human primate models. In the first, KPL-716 abrogated the pharmacodynamic marker of pruritus in an IL-31 challenge model. A single three milligram per kilogram dose of KPL-716 substantially reduced scratch counts despite multiple repeated injections of IL-31 over

several weeks at concentrations we believe to be supraphysiologic in a disease context. In the second non-human primate model, KPL-716 again abrogated the painful response to an injection to an inflammatory agent called carrageenan through the time period measured after a single infusion of KPL-716, implicating OSM in the inflammatory response. We have conducted pre-clinical toxicology studies for KPL-716 with a no adverse event level of 500 milligrams per kilogram with intravenous dosing.

Phase 1a/1b Clinical Trial

In early 2017, we filed an IND application and began clinical development with KPL-716 in a Phase 1a/1b clinical trial in healthy volunteers and in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus, respectively. The trial uses a double-blind, randomized, placebo-controlled, sequential-group design to evaluate the safety, tolerability, pharmacokinetics and immunogenicity of KPL-716 in male and female subjects. In addition, in the single-dose Phase 1b portion of the trial, the cohorts of subjects with moderate-to-severe atopic dermatitis received dose levels of KPL-716 that could show an early signal of efficacy in reducing pruritus as an exploratory endpoint using a validated Numerical Rating Score which measures pruritus intensity. We have completed dosing in the first portion of the Phase 1a/1b clinical trial, which is a single ascending dose design. Each dosing cohort received a single dose of KPL-716 administered intravenously or subcutaneously. In the second portion of the Phase 1b clinical trial, each subject will receive repeated single doses of KPL-716 administered subcutaneously. We expect to report preliminary data from the single ascending dose cohorts of this portion of this trial in the second half of 2018. In the second portion of the Phase 1a/1b clinical trial, each subject in the cohort will receive repeated single doses of KPL-716 administered subcutaneously. We plan to commence dosing in this portion of the trial in 2018, subject to submission to, and review by, the FDA and Canadian regulatory authorities of our recently completed primate chronic toxicology study.

We are also conducting an observational study in prurigo nodularis patients called LOTUS-PN. Our LOTUS-PN study will explore the extent to which clinical endpoints correlate with mechanistic biomarkers. In consideration of the importance of humanistic parameters, the LOTUS-PN study will also examine the impact of prurigo nodularis and various treatment options on quality of life. Ultimately, the LOTUS-PN study will seek to provide a better understanding of the clinical presentation and course of prurigo nodularis: its pathogenesis, treatment regimens, outcomes and inter- and intra-patient variability.

Pre-clinical Development

KPL-045

KPL-045 is a fully-human monoclonal antibody that is designed to inhibit the CD30-CD30 ligand interaction, a co-stimulatory signal helpful in activating and sustaining memory T-cells. The majority of the therapeutics in development modulating the CD30-CD30 ligand interaction are depleting or conjugated to a toxin for the use in hematological malignancies. To our knowledge, KPL-045 is the only non-depleting antibody targeting primarily autoimmune disease in active clinical development. In August 2017, we licensed this antibody from Novo Nordisk.

In pre-clinical development, of KPL-045 has been observed to have a favorable pharmacokinetic profile to support further development. KPL-045 has demonstrated single-digit nanomolar potency against both human and cynomolgus non-human primate CD30L. We are planning IND-enabling studies in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in 2019.

KPL-404

KPL-404 is a humanized monoclonal antibody that is designed to inhibit the CD40-CD40 ligand interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching. We have a license to conduct research and development on KPL-404 from Primatope Therapeutics, Inc., or Primatope, the company that owns or controls the intellectual property related to KPL-404. We also have an exclusive option to acquire all outstanding capital stock of Primatope, which, subject to extension, is exercisable until January 2019.

In pre-clinical development, KPL-404 has been observed to have a favorable pharmacokinetic and toxicology profile to support further development. KPL-404 has been effective in multiple non-human primate models of organ transplant rejection, as well as in multiple T-cell dependent antibody response models. We are planning IND-enabling studies in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in 2019.

Discovery Activities

We have initiated internal discovery activities directed toward wholly owned molecules for the treatment of autoinflammatory and autoimmune disease targets where we believe there to be a strong mechanistic rationale and clear differentiation from existing approved agents or those in development.

License and Acquisition Agreements

License Agreement with Regeneron

In September 2017, we entered into a license agreement with Regeneron, or the Regeneron Agreement. Pursuant to the Regeneron Agreement, Regeneron granted us an exclusive license under certain intellectual property rights controlled by Regeneron to develop and commercialize rilonacept worldwide, aside from Israel, Egypt, Turkey and select countries in the Middle East and northern Africa, which we refer to collectively as the Excluded Territory. In the United States and Japan, our license is initially for all indications other than those involving local administration to the eye or ear, oncology, deficiency of the interleukin-1 receptor antagonist, or DIRA, and CAPS. If we are successful in receiving marketing approval for rilonacept in the United States for a new indication, the scope of the license granted to us will automatically expand to include DIRA and CAPS in the United States and Japan, and we will assume the sales and distribution of rilonacept in these additional indications. Outside the U.S. and Japan, our license is for all indications other than local application to the eye or ear, oncology, CAPS, DIRA and certain periodic fever syndromes set forth in the Regeneron Agreement, collectively the Excluded Indications. Under the Regeneron Agreement, we are obligated to use commercially reasonable efforts to develop and commercialize rilonacept outside of the Excluded Indications in our territory. Upon receiving positive data in a Phase 3 clinical trial, Regeneron will transfer the BLA for rilonacept to us.

We made an upfront payment of $5.0 million to Regeneron and are obligated to make regulatory milestone payments of up to $27.5 million in the aggregate. Thereafter, we have agreed to evenly split profits on our sales of rilonacept with Regeneron after deducting certain commercialization expenses subject to specified limits.

Regeneron has a right of first negotiation over our engagement of third parties to support our promotional activities in excess of a specified level and over the assignment or sale of our rights to any product we develop under the Regeneron Agreement to a third-party. Furthermore under certain circumstances, we will need Regeneron's prior consent to assign our rights under the Regeneron Agreement.

The Regeneron Agreement will expire on the date on which we, our affiliates or sublicensees are no longer developing or commercializing any product containing rilonacept. We may terminate

the agreement for convenience at any time after the date that is 18 months after the effective date of the agreement with 180 days' written notice or one year's written notice if we terminate the agreement following U.S. marketing approval of a rilonacept product developed by us. We may also terminate with three months' written notice if we reasonably determine that rilonacept is unsafe in the indications we are pursuing. Regeneron may terminate the agreement if there is a consecutive twelve (12) month period during which we do not conduct any material development or commercialization activities or we do not grant a sublicense to a third-party to do so, or if we challenge Regeneron's patent rights in any country in our territory. Either party may terminate the agreement in the event of a material breach by the other party that remains uncured for 90 days (or 30 days for payment-related breaches), or by either party due to the insolvency or bankruptcy of the other party.

We have also entered into a clinical supply agreement with Regeneron, or the Supply Agreement. Pursuant to the Supply Agreement, Regeneron has the exclusive right to manufacture and supply all of our requirements of rilonacept for clinical development. If Regeneron determines to discontinue the supply of rilonacept to us, it must use its reasonable efforts to transfer all relevant documentation, materials and technology necessary for the manufacture of rilonacept to us or our designee. The Supply Agreement terminates upon the termination of the Regeneron Agreement or the transfer of technology related to the bulk manufacture of rilonacept.

License Agreement with MedImmune

In December 2017, we entered into a license agreement with MedImmune, or the MedImmune Agreement. Pursuant to the MedImmune Agreement, MedImmune granted us an exclusive, worldwide license under certain intellectual property rights controlled by MedImmune to make, use, develop and commercialize mavrilimumab and any other product containing an antibody to the GM-CSF receptor alpha that is covered by certain MedImmune patent rights for all indications. We also acquired non-exclusive licenses to other MedImmune technology for use in exploiting licensed products. We may sublicense these rights subject to consent of MedImmune and any applicable licensors of rights under which we are licensed. We also acquired reference rights to relevant manufacturing and regulatory documents, and existing inventory of mavrilimumab drug substance. We must use commercially reasonable efforts to develop and commercialize the licensed products.

We made an upfront payment of $8.0 million to MedImmune and are obligated to make future clinical, regulatory and initial sales milestone payments of up to $72.5 million in the aggregate for the first two indications, including a milestone payment of $10.0 million upon the earlier to occur of a specified regulatory milestone and December 31, 2018, and clinical and regulatory milestone payments of up to $15.0 million in the aggregate for each subsequent indication. We are also obligated to make milestone payments to MedImmune of up to $85.0 million upon the achievement of annual net sales thresholds up to, but excluding, $1.0 billion in annual net sales as well as additional milestone payments aggregating up to $1.1 billion upon the achievement of additional annual net sales thresholds starting at $1.0 billion and higher. Commencing on the first commercial sale of licensed products, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. We must pay such royalties on a product-by-product and country-by-country basis until the latest to occur of the expiration of licensed patents, the expiration of regulatory exclusivity or the tenth anniversary of first commercial sale of such product in such country.

In countries where licensed patents have issued, the statutory expiration date is 2027, not including any patent term extensions or adjustments. While the current expected patent expiration dates are known in countries where licensed patents have issued, these expiration dates are subject to significant uncertainty. For example, the patents may be challenged, and accordingly, the relevant expiration dates could be shortened. In addition, as we continue to file and prosecute patent applications related to mavrilimumab, the granting of pending applications or future patent

applications could extend the relevant statutory expiration dates beyond 2027. The expiration date of regulatory exclusivity is determined on a country-by country-basis if the applicable product is approved in such country and if any applicable regulatory exclusivity applies and is granted. The actual expiration date of any such regulatory exclusivity, however, is subject to significant uncertainty. For instance, the applicable regulatory exclusivity period is often triggered by the date a product candidate obtains regulatory approval, and we cannot predict with any certainty whether and if so, when, the applicable product would receive regulatory approval in any given jurisdiction. Furthermore, the type, scope and duration of such exclusivities will vary on a country-by-country basis depending on the jurisdictions in which a product candidate is approved and the particular regulatory exclusivity for which the product is eligible as of the time of approval. For example, in the United States, a reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, which means that the FDA cannot make effective the approval of a biosimilar product that references the biologic product until 12 years from the date on which the reference product was first licensed. In the European Union, new products authorized for marketing may qualify for eight years of data exclusivity and an additional two years of market exclusivity upon marketing authorization. The two-year period may be extended to three years if during the first eight years a new therapeutic indication with significant clinical benefit over existing therapies is approved. Furthermore, if a product candidate that has received orphan designation is subsequently approved for the disease or condition for which it has such designation, the product may be entitled to orphan drug exclusivity, which generally grants seven years of market exclusivity in the United States and up to 10 years of market exclusivity in the European Union, and such period may run contemporaneously with the other exclusivities that may apply. In the European Union, an orphan product can also obtain an additional two years of market exclusivity for pediatric studies. In the United States, an additional six-month period of pediatric exclusivity may be available as an extension to any existing non-patent regulatory exclusivity period if the sponsor has conducted and submitted pediatric studies in response to a written request from the FDA. Additionally, our eligibility for regulatory exclusivity may depend in part on the indications for which we seek regulatory approval of our product candidates, which may depend on the data we receive from our clinical studies, and accordingly, may change over time, and the laws and regulations governing regulatory exclusivity may change in various jurisdictions as the political focus on drug exclusivity increases. For risk related to regulatory exclusivity matters, see "Risk Factors—Risks Related to Product Development and Regulatory Approval."

The MedImmune Agreement will remain in effect until the expiration of the royalty term in all countries for all licensed products. The MedImmune Agreement may be terminated earlier at any time by us with at least 90 days' prior notice, by either party in the event of material breach by the other party that remains uncured for 90 days, by either party for insolvency or bankruptcy of the other party, or immediately by MedImmune if we challenge the licensed patents.

Biogen Asset Purchase Agreement

In September 2016, we completed the acquisition of certain assets of Biogen pursuant to an asset purchase agreement, or the Biogen Agreement. Pursuant to the Biogen Agreement, we acquired all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716 and other antibodies covered by certain patent rights, together the Acquired Assets, including patents and other intellectual property rights, clinical data, certain contracts, know-how and inventory. In addition, Biogen granted us a non-exclusive, sublicensable, worldwide license to certain background patent rights related the KPL-716 program. Under the Biogen Agreement, we are obligated to use commercially reasonable efforts to develop and commercialize the Acquired Assets.

Under the Biogen Agreement, we made an upfront payment of $11.5 million and a technology transfer payment of $0.5 million to Biogen. In addition, we made a milestone payment of

$4.0 million during the year ended December 31, 2017 associated with the achievement of a specified clinical milestone event. We are also obligated to make future milestone payments for each antibody product that includes the Acquired Assets, or an Antibody Product, of up to $325.0 million in the aggregate upon the achievement of specified milestones. These milestone payments relate to multiple indications for an Antibody Product, and are comprised of up to $175.0 million in the aggregate upon achievement of specified clinical and regulatory milestone events and $150.0 million in the aggregate upon the achievement of specified annual net sales thresholds. Commencing on the first commercial sale of an Antibody Product, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens. We must pay such royalties on a product-by-product and country-by-country basis until the latest to occur of the expiration of patents that cover an Antibody Product, the expiration of regulatory exclusivity or the tenth anniversary of first commercial sale of such product in such country. We have also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to KPL-716.

In countries where patents covering Antibody Products have issued, the statutory expiration date is 2034, not including any patent term extensions or adjustments. While the current expected patent expiration dates are known in countries where licensed patents have issued, these expiration dates are subject to significant uncertainty. For example, the patents may be challenged, and accordingly, the relevant expiration dates could be shortened. In addition, as we continue to file and prosecute patent applications related to Antibody Products, the granting of pending applications or future patent applications could extend the relevant statutory expiration dates beyond 2034. The expiration date of regulatory exclusivity is determined on a country-by country-basis if the applicable product is approved in such country and if any applicable regulatory exclusivity applies and is granted. The actual expiration date of any such regulatory exclusivity, however, is subject to significant uncertainty. For instance, the applicable regulatory exclusivity period is often triggered by the date a product candidate obtains regulatory approval, and we cannot predict with any certainty whether and if so, when, the applicable product would receive regulatory approval in any given jurisdiction. Furthermore, the type, scope and duration of such exclusivities will vary on a country-by-country basis depending on the jurisdictions in which a product candidate is approved and the particular regulatory exclusivity for which the product is eligible as of the time of approval. For example, in the United States, a reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, which means that the FDA cannot make effective the approval of a biosimilar product that references the biologic product until 12 years from the date on which the reference product was first licensed. In the European Union, new products authorized for marketing may qualify for eight years of data exclusivity and an additional two years of market exclusivity upon marketing authorization. The two-year period may be extended to three years if during the first eight years a new therapeutic indication with significant clinical benefit over existing therapies is approved. Furthermore, if a product candidate that has received orphan designation is subsequently approved for the disease or condition for which it has such designation, the product may be entitled to orphan drug exclusivity, which generally grants seven years of market exclusivity in the United States and up to 10 years of market exclusivity in the European Union, and such period may run contemporaneously with the other exclusivities that may apply. In the European Union, an orphan product can also obtain an additional two years of market exclusivity for pediatric studies. In the United States, an additional six-month period of pediatric exclusivity may be available as an extension to any existing non-patent regulatory exclusivity period if the sponsor has conducted and submitted pediatric studies in response to a written request from the FDA. Additionally, our eligibility for regulatory exclusivity may depend in part on the indications for which we seek regulatory approval of our product candidates, which may depend on the data we receive from our clinical studies, and accordingly, may change over time, and the laws and regulations governing regulatory exclusivity may change in various jurisdictions as the political

focus on drug exclusivity increases. For risk related to regulatory exclusivity matters, see "Risk Factors—Risks Related to Product Development and Regulatory Approval."

Under the Biogen Agreement, Biogen has a time-limited right of first negotiation to purchase the assets we acquired from Biogen or obtain a license to exploit Antibody Products, in each case, in the event we decide to sell the acquired assets, including through the sale of our company, or out-license the rights to the Antibody Products.

The Biogen Agreement will remain in effect until expiration of all payment obligations in all countries related to the last antibody product subject to the Biogen Agreement. The Biogen Agreement may be terminated by us with 90 days' prior notice, by either party in the event of a material breach by the other party that remains uncured for 90 days (or 30 days for payment-related breaches) or by both parties upon mutual consent. In the event of a termination, the Acquired Assets, including certain licenses and rights related thereto, will revert to Biogen, and, upon written request by Biogen, we are required to grant to Biogen an exclusive, worldwide, sub-licensable license to certain of our intellectual property related to the Acquired Assets, including know-how and patent rights.

Manufacturing

We rely on third parties to manufacture all of our product candidates. We have entered into a clinical supply agreement with Regeneron to manufacture and supply rilonacept for our clinical trials. Regeneron has also agreed to provide commercial drug material until at least the later of four years after U.S. marketing approval or seven years after the effective date of the agreement.

We believe that we have sufficient quantities of drug substance to supply our planned Phase 2 clinical trial of mavrilimumab for the treatment of GCA. We also acquired a certain amount of finished mavrilimumab drug product that we plan to use in this clinical trial, and we intend to enter into a fill/finish supply agreement with a contract manufacturing organization, or CMO, to produce additional finished mavrilimumab drug product from our current inventory of drug substance. We plan to identify and enter into an agreement with a CMO to produce mavrilimumab drug substance beyond our existing inventory for any further clinical trials and eventual commercialization of mavrilimumab, if approved. There are certain components, for example, media and feed, used to produce our current mavrilimumab inventory that we will not use in our future manufacturing process. We and any CMO that we enter into agreement with to manufacture mavrilimumab will need to find alternative components to replace the media and feed that had been used by MedImmune to date in the manufacture of mavrilimumab.

We acquired a certain amount of KPL-716 drug substance from Biogen from which we produced KPL-716 drug product using a CMO. In addition, we have engaged CMOs to manufacture KPL-716 drug substance and product for further clinical development activities. We intend to continue using CMOs to develop our manufacturing process and scale-up for any future clinical trials and eventual commercialization of KPL-716, if approved.

We are using CMOs to produce our pre-clinical product candidates, KPL-045 and KPL-404, for our planned IND-enabling studies.

We require all of our CMOs to conduct manufacturing activities in compliance with current good manufacturing practice, or cGMP, requirements. We have assembled a team of experienced employees and consultants to provide the necessary technical, quality and regulatory oversight of our CMOs. We currently rely solely on these third-party manufacturers for scale-up and process development work and to produce sufficient quantities of product candidate for use in pre-clinical studies and clinical trials. Although we have plans to establish our own manufacturing capabilities to support certain pre-clinical and early clinical-stage production of product candidates, we intend to continue to rely on third-party manufacturers for clinical and commercial supply of our product

candidates. We anticipate that these CMOs will have the capacity to support both clinical supply and commercial-scale production, but we do not have any formal agreements at this time with any of these CMOs to cover commercial production. We also may elect to pursue additional CMOs for manufacturing supplies of drug substance and finished drug product in the future.

Commercial Operations

Our team is experienced in commercial leadership and we intend to expand our capabilities in parallel with the development path of our product candidates. If the FDA approves rilonacept for recurrent pericarditis, we intend to market and commercialize rilonacept in the United States by developing our own sales, marketing and medical affairs organizations targeting a subset of cardiologists and rheumatologists currently treating pericarditis. For our other product candidates, we intend to establish commercialization strategies for each as we approach potential marketing approval and, due to the specialization among physicians treating the indications we are targeting, we expect to be able leverage our then-existing sales, marketing and medical affairs organizations.

Competition

The biotechnology and pharmaceutical industries are characterized by rapidly advancing technologies, intense competition and a strong emphasis on proprietary products. We face potential competition from many different sources, including pharmaceutical and biotechnology companies, academic institutions and governmental agencies and public and private research institutions. Any product candidates that we successfully develop and commercialize, including rilonacept, mavrilimumab and KPL-716, may compete with existing products and new products that may become available in the future.

Our competitors may have significantly greater financial resources, established presence in the market, expertise in research and development, manufacturing, pre-clinical and clinical testing, obtaining regulatory approvals and reimbursement and marketing approved products than we do. These competitors also compete with us in recruiting and retaining qualified scientific, sales, marketing and management personnel, establishing clinical trial sites and patient registration for clinical trials, as well as in acquiring technologies complementary to, or necessary for, our programs. Smaller or early-stage companies may also prove to be significant competitors, particularly through collaborative arrangements with large and established companies.

The key competitive factors affecting the success of rilonacept, mavrilimumab and KPL-716, and any other product candidates that we develop, if approved, are likely to be their efficacy, safety, convenience, price, the level of generic competition and the availability of reimbursement from government and other third-party payors. Our commercial opportunity for any of our product candidates could be reduced or eliminated if our competitors develop and commercialize products that are more effective, have fewer or less severe side effects, are more convenient or are less expensive than any products that we may develop. Our competitors also may obtain FDA or other regulatory approval for their products more rapidly than we may obtain approval for ours. In addition, our ability to compete may be affected in many cases by insurers or other third-party payors seeking to encourage the use of generic products.

We are aware of the following products currently marketed or in clinical development for the treatment of the diseases that we are initially targeting:

Rilonacept

We are not aware of any therapies currently approved by the FDA for the treatment of recurrent pericarditis, our lead indication for rilonacept. Anakina (KINERET), produced by Sobi, Inc., is an FDA-approved agent that inhibits IL-1a and IL-1b signaling and is approved for RA and CAPS. Canakinumab (ILARIS), produced by Novartis Pharmaceuticals Corporation, is a monoclonal

antibody which inhibits IL-1b signaling and is approved for use in CAPS, tumor necrosis factor receptor associated period syndrome, hyperimmunoglobulin D syndrome, familiar Mediterranean fever and active systemic juvenile idiopathic arthritis. There are also other therapies modulating IL-1a and/or IL-1b which are in various stages of clinical development for diseases other than recurrent pericarditis from AbbVie, Inc., or AbbVie, XBiotech Inc. and Handok Inc.

Mavrilimumab

Tocilizumab (ACTEMRA), produced by Hoffmann — La Roche AG, or Roche, and Chugai Pharmaceutical Co., Ltd., is an IL-6 inhibitor that is approved by the FDA for the treatment of GCA on top of a concomitant corticosteroid taper. There are also four other programs in clinical development that modulate GM-CSF signaling from GlaxoSmithKline plc, or GSK, Izana Bioscience Ltd., Morphotek, Inc. and Humanigen, Inc. In addition, Eli Lilly and AbbVie are conducting clinical trials for oral janus kinase inhibitors, and Sanofi S.A. and Regeneron intend to initiate a Phase 3 clinical trial with their anti-IL-6 program in 2018.

KPL-716

We are not aware of any therapies currently approved by the FDA for the treatment of prurigo nodularis. Menlo Therapeutics Inc., Vanda Pharmaceuticals Inc., Trevi Therapeutics, Inc. and Galderma SA, or Galderma, have programs in various stages of clinical development for the treatment of prurigo nodularis.

The FDA recently approved Regeneron's dupilumab, an antibody that inhibits signaling through the interleukin 4 receptor, to treat atopic dermatitis. Other companies currently developing systemic therapies for atopic dermatitis include Roche, Dermira, Inc., Galderma, Asana BioSciences, LLC, Eli Lilly and Co., Pfizer Inc., AbbVie, Glenmark Pharmaceuticals Ltd., GSK, LEO Pharma Inc., Incyte Corporation, Dermavant Sciences, Inc. and AnaptsysBio, Inc.

Intellectual Property

Our success depends in part on our ability to obtain and maintain proprietary protection for our drug candidates, manufacturing and process discoveries, and other know-how, to operate without infringing the proprietary rights of others, and to prevent others from infringing our proprietary rights. We plan to protect our proprietary position using a variety of methods, which include pursuing U.S. and foreign patent applications related to our proprietary technology, inventions and improvements, including compositions of matter and methods-of-use, that are important to the development and implementation of our business. For example, we or our licensors have or are pursuing patents covering the composition of matter for each of our product candidates and we generally pursue patent protection covering methods-of-use for each clinical program. We also rely on trade secrets, know-how, continuing technological innovation and potential in-licensing opportunities to develop and maintain our proprietary position.

We have a field-specific exclusive license under the Regeneron Agreement to granted patents and pending applications in the United States and numerous foreign jurisdictions relating to rilonacept. As of March 31, 2018, the patent rights in-licensed under the Regeneron Agreement relating to our program include one granted patent in the United States and 53 patents granted in foreign jurisdictions, including Canada, Australia, Brazil and selected countries in Europe and Asia. In addition, the patent rights in-licensed under the Regeneron Agreement relating to our program include patent applications that are pending in the United States. A U.S. patent covering rilonacept as a composition of matter has a statutory expiration date in 2019, not including patent term adjustment, and relevant foreign counterparts are expected to expire between 2019 and 2023, in each case, not including any patent term extensions. If we are successful in obtaining regulatory approval of rilonacept for the treatment of recurrent pericarditis, we expect to rely on orphan

exclusivity, which generally grants seven years of marketing exclusivity in the United States and 10 years of marketing exclusivity in Europe. See "—License Agreement with Regeneron" above for additional information on our rights under the Regeneron Agreement.

We have an exclusive license under the MedImmune Agreement to granted patents and pending patent applications in the United States and numerous foreign jurisdictions relating to mavrilimumab. These patents and patent applications cover mavrilimumab as a composition of matter and its use. As of March 31, 2018, the patent rights in-licensed under the MedImmune Agreement relating to our program include three granted patents in the United States and 105 patents granted in foreign jurisdictions, including Canada, Australia and selected countries in Europe and Asia. In addition, the patent rights in-licensed under the MedImmune Agreement relating to our program include patent applications that are pending in the United States and selected countries in Asia and Latin America. The composition of matter patents for mavrilimumab generally have statutory expiration dates in 2027, although the term of some U.S. patents may be longer due to patent term adjustment to compensate for delays during the patent prosecution process. Patent term extension could extend the expiration date of one patent in the United States and patents in certain other jurisdictions, each in accordance with applicable law. There can be no assurances that patents will issue from any pending patent applications. See "—License Agreement with MedImmune" above for additional information on our rights under the MedImmune Agreement.

We own, via our acquisition of certain assets from Biogen, granted patents and pending patent applications in the United States and numerous foreign jurisdictions relating to KPL-716. These patents and patent applications cover KPL-716 as a composition of matter and its use. As of March 31, 2018, the patent rights acquired from Biogen include three patents granted in the United States and four foreign patents granted in Australia, Japan, Mexico and Singapore. In addition, the patent rights acquired from Biogen include patent applications pending in the United States, Europe, Canada, and selected countries in Asia. The issued composition of matter patents for KPL-716 have statutory expiration dates in 2034. Patent term extension could extend the expiration date of one patent in the United States and patents in certain other jurisdictions, each in accordance with applicable law. There can be no assurance that patents will issue from any of our pending patent applications. See "—Biogen Asset Purchase Agreement" above for additional information on our rights under the Biogen Agreement.

The term of individual patents depends upon the legal term for patents in the countries in which they are obtained. In most countries, including the United States, the patent term is 20 years from the earliest filing date of a non-provisional patent application. In the United States, a patent's term may be lengthened by patent term adjustment, which compensates a patentee for administrative delays by the USPTO, in examining and granting a patent, or may be shortened if a patent is terminally disclaimed over an earlier expiring patent. In certain countries, the term of a patent that covers a drug product may also be eligible for patent term extension when regulatory approval is granted, provided the legal requirements are met. In the future, if and when our drug candidates receive approval by the FDA or foreign regulatory authorities, we expect to apply for patent term extensions on issued patents covering those drugs, depending upon the length of the clinical trials for each drug and other factors. There can be no assurance that any of our pending patent applications will issue or that we will benefit from any patent term extension or favorable adjustment to the term of any of our patents.

Government Regulation

Government authorities in the United States at the federal, state and local level and in other countries and jurisdictions, including the European Union, extensively regulate, among other things, the research, development, testing, manufacture, quality control, approval, labeling, packaging, storage, record-keeping, promotion, advertising, distribution, post-approval monitoring and reporting, marketing and export and import of drug products, such as rilonacept, mavrilimumab and

our other product candidates. Generally, before a new drug can be marketed, considerable data demonstrating its quality, safety and efficacy must be obtained, organized into a format specific for each regulatory authority, submitted for review and approved by the regulatory authority.

U.S. Government Regulation of Biological Products

In the United States, the FDA regulates biologics under the Federal Food, Drug, and Cosmetic Act, or FDCA, and the Public Health Service Act, or PHSA, and their implementing regulations. Products are also subject to other federal, state and local statutes and regulations. The process of obtaining regulatory approvals and the subsequent compliance with applicable federal, state, local and foreign statutes and regulations requires the expenditure of substantial time and financial resources. Failure to comply with the applicable U.S. requirements at any time during the product development process, approval process or after approval, may subject an applicant to a variety of administrative or judicial sanctions, such as the FDA's refusal to approve pending biologic license applications, or BLAs, withdrawal of an approval, imposition of a clinical hold, issuance of warning letters, product recalls, product seizures, total or partial suspension of production or distribution, injunctions, fines, refusals of government contracts, restitution, disgorgement of profits or civil or criminal penalties.

The process required by the FDA before a biologic may be marketed in the United States generally involves the following:

- Completion of extensive pre-clinical studies and tests in accordance with applicable regulations, including Good Laboratory Practice, or GLP, regulations and applicable requirements for the humane use of laboratory animals or other applicable regulations;
- Submission to FDA of an IND which must become effective before human clinical trials may begin;
- Approval by an independent institutional review board, or IRB, or ethics committee at each clinical trial site before each trial may be initiated;
- Performance of adequate and well-controlled human clinical trials in accordance with applicable IND regulations, good clinical practices, or GCPs, and other clinical-trial related regulations to evaluate the safety and efficacy of the investigational product for each proposed indication;
- Submission to FDA of a BLA for marketing approval that includes substantive evidence of safety, purity, and potency from results of pre-clinical testing and clinical trials;
- A determination by FDA within 60 days of its receipt of a BLA to accept the filing for review;
- Satisfactory completion of one or more FDA pre-approval inspections of the manufacturing facility or facilities where the biologic will be produced to assess compliance with cGMPs to assure that the facilities, methods and controls used in product manufacture are adequate to preserve the biologic's identity, strength, quality and purity;
- Potential FDA audit of the pre-clinical and/or clinical trial sites that generated the data in support of the BLA;
- Payment of user fees for FDA review of the BLA; and
- FDA review and approval of the BLA, including satisfactory completion of an FDA advisory committee review, if applicable, prior to any commercial marketing or sale of the product in the United States.

Pre-clinical Studies

Before testing any biological product candidate, including our product candidates, in humans, the product candidate must undergo rigorous pre-clinical testing. The pre-clinical development stage generally involves laboratory evaluations of the chemistry, formulation and stability of the product candidate, as well as trials to evaluate toxicity in animals, which support subsequent clinical testing. The conduct of the pre-clinical tests must comply with federal regulations and requirements, including GLP regulations. The sponsor must submit the results of the pre-clinical studies, together with manufacturing information, analytical data, any available clinical data or literature and a proposed clinical protocol, to the FDA as part of the IND. An IND is a request for authorization from the FDA to administer an investigational product to humans, and must become effective before human clinical trials may begin. Some pre-clinical testing may continue even after the IND is submitted. An IND automatically becomes effective 30 days after receipt by the FDA, unless before that time the FDA raises concerns or questions related to one or more proposed clinical trials and places the clinical trial on a clinical hold. In such a case, the IND sponsor and the FDA must resolve any outstanding concerns before the clinical trial can begin. As a result, submission of an IND may not result in the FDA allowing clinical trials to commence.

Clinical Trials

The clinical stage of development involves the administration of the investigational product to healthy volunteers or patients under the supervision of qualified investigators, generally physicians not employed by, or under control of, the trial sponsor, in accordance with GCPs, which include the requirement that all research subjects provide their informed consent for their participation in any clinical trial. Clinical trials are conducted under protocols detailing, among other things, the objectives of the clinical trial, dosing procedures, subject selection and exclusion criteria and the parameters to be used to monitor subject safety and assess efficacy. Each protocol, and any subsequent amendments to the protocol, must be submitted to the FDA as part of the IND. Furthermore, each clinical trial must be reviewed and approved by an IRB for each institution at which the clinical trial will be conducted to ensure that the risks to individuals participating in the clinical trials are minimized and are reasonable in relation to anticipated benefits. The IRB also approves the informed consent form that must be provided to each clinical trial subject or his or her legal representative, and must monitor the clinical trial until completed. There also are requirements governing the reporting of ongoing clinical trials and completed clinical trial results to public registries. Information about most clinical trials must be submitted within specific timeframes for publication on the *www.clinicaltrials.gov* website. Information related to the product, patient population, phase of investigation, study sites and investigators and other aspects of the clinical trial is made public as part of the registration of the clinical trial. Sponsors are also obligated to discuss the results of their clinical trials after completion. Disclosure of the results of these trials can be delayed in some cases for up to two years after the date of completion of the trial. Competitors may use this publicly available information to gain knowledge regarding the progress of development programs.

Human clinical trials are typically conducted in three sequential phases, which may overlap or be combined:

- Phase 1 clinical trials generally involve a small number of healthy volunteers or disease-affected patients who are initially exposed to a single dose and then multiple doses of the product candidate. The primary purpose of these clinical trials is to assess the metabolism, pharmacologic action, side effect tolerability and safety of the drug.

- Phase 2 clinical trials involve studies in disease-affected patients to determine the dose required to produce the desired benefits. At the same time, safety and further

pharmacokinetic and pharmacodynamic information is collected, possible adverse effects and safety risks are identified and a preliminary evaluation of efficacy is conducted.

- Phase 3 clinical trials generally involve a larger number of patients at multiple sites and are designed to provide the data necessary to demonstrate the effectiveness of the product for its intended use, its safety in use and to establish the overall benefit/risk relationship of the product and provide an adequate basis for product approval. These trials may include comparisons with placebo and/or other comparator treatments. The duration of treatment is often extended to mimic the actual use of a product during marketing.

Post-approval trials, sometimes referred to as Phase 4 clinical trials, may be conducted after initial marketing approval. These trials are used to gain additional experience from the treatment of patients in the intended therapeutic indication, particularly for long-term safety follow up. In certain instances, the FDA may mandate the performance of Phase 4 clinical trials as a condition of approval of a BLA.

Progress reports detailing the results of the clinical trials must be submitted at least annually to the FDA and more frequently if serious adverse events occur. The FDA or the sponsor may suspend or terminate a clinical trial at any time or the FDA may impose other sanctions on various grounds, including a finding that the research patients are being exposed to an unacceptable health risk. Similarly, an IRB can suspend or terminate approval of a clinical trial at its institution if the clinical trial is not being conducted in accordance with the IRBs requirements or if the drug has been associated with unexpected serious harm to patients.

Concurrently with clinical trials, companies usually complete additional pre-clinical studies and must also develop additional information about the physical characteristics of the biological product as well as finalize a process for manufacturing the product in commercial quantities in accordance with cGMP requirements. To help reduce the risk of the introduction of adventitious agents with use of biological products, the PHSA emphasizes the importance of manufacturing control for products whose attributes cannot be precisely defined. The manufacturing process must be capable of consistently producing quality batches of the product candidate and, among other things, the sponsor must develop methods for testing the identity, strength, quality, potency and purity of the final biological product. Additionally, appropriate packaging must be selected and tested and stability studies must be conducted to demonstrate that the biological product candidate does not undergo unacceptable deterioration over its shelf life.

BLA Review and Approval

Assuming successful completion of the required clinical testing, the results of the pre-clinical studies and clinical trials, together with detailed information relating to the product's chemistry, manufacture, controls and proposed labeling, among other things, are submitted to the FDA as part of a BLA requesting approval to market the product for one or more indications. The BLA must contain proof of safety, purity, potency and efficacy and may include both negative and ambiguous results of pre-clinical studies and clinical trials as well as positive findings. Data may come from company-sponsored clinical trials intended to test the safety and efficacy of a product's use or from a number of alternative sources, including studies initiated by investigators. To support marketing approval, the data submitted must be sufficient in quality and quantity to establish the safety and efficacy of the investigational product to the satisfaction of the FDA. FDA approval of a BLA must be obtained before a biologic may be marketed in the United States.

In most cases, the submission of a BLA is subject to a substantial application user fee. Under the goals and policies agreed to by the FDA under the Prescription Drug User Fee Act, or PDUFA, for original BLAs, the FDA has ten months from the filing date in which to complete its initial review of a standard application and respond to the applicant, and six months from the filing date for an

application with priority review. The FDA does not always meet its PDUFA goal dates, and the review process is often significantly extended by FDA requests for additional information or clarification. This review typically takes twelve months from the date the BLA is submitted to the FDA because the FDA has approximately two months to make a "filing" decision.

Before approving a BLA, the FDA will typically conduct a pre-approval inspection of the manufacturing facilities for the new product to determine whether the manufacturing processes and facilities comply with cGMPs. The FDA will not approve the product unless it determines that the manufacturing processes and facilities are in compliance with cGMP requirements and adequate to assure consistent production of the product within required specifications. The FDA also may inspect the sponsor and one or more clinical trial sites to assure compliance with GCP requirements and the integrity of the clinical data submitted to the FDA.

Additionally, the FDA may refer applications for novel biologic candidates which present difficult questions of safety or efficacy to an advisory committee, typically a panel that includes clinicians and other experts, for review, evaluation and a recommendation as to whether the application should be approved and under what conditions, if any. The FDA is not bound by recommendations of an advisory committee, but it considers such recommendations when making decisions on approval. The FDA likely will re-analyze the clinical trial data, which could result in extensive discussions between the FDA and the applicant during the review process. The FDA also may require submission of a REMS plan if it determines that a REMS is necessary to ensure that the benefits of the drug outweigh its risks and to assure the safe use of the biological product. The REMS plan could include medication guides, physician communication plans, assessment plans and/or elements to assure safe use, such as restricted distribution methods, patient registries or other risk minimization tools. The FDA determines the requirement for a REMS, as well as the specific REMS provisions, on a case-by-case basis. If the FDA concludes a REMS is needed, the sponsor of the BLA must submit a proposed REMS. The FDA will not approve a BLA without a REMS, if required.

Under the Pediatric Research Equity Act, or PREA, a BLA or supplement to a BLA must contain data that are adequate to assess the safety and efficacy of the product candidate for the claimed indications in all relevant pediatric populations and to support dosing and administration for each pediatric population for which the product is safe and effective. The FDA may grant deferrals for submission of pediatric data or full or partial waivers. The Food and Drug Administration Safety and Innovation Act, or FDASIA, amended the FDCA to require that a sponsor who is planning to submit a marketing application for a product that includes a new active ingredient, new indication, new dosage form, new dosing regimen or new route of administration submit an initial Pediatric Study Plan, or PSP, within sixty days of an end-of-Phase 2 meeting or, if there is no such meeting, as early as practicable before the initiation of the Phase 3 or Phase 2/3 clinical trial. The initial PSP must include an outline of the pediatric study or studies that the sponsor plans to conduct, including trial objectives and design, age groups, relevant endpoints and statistical approach, or a justification for not including such detailed information, and any request for a deferral of pediatric assessments or a full or partial waiver of the requirement to provide data from pediatric studies along with supporting information. The FDA and the sponsor must reach an agreement on the PSP. A sponsor can submit amendments to an agreed upon initial PSP at any time if changes to the pediatric plan need to be considered based on data collected from pre-clinical studies, early phase clinical trials or other clinical development programs.

After the FDA evaluates a BLA, it will issue an approval letter or a Complete Response Letter. An approval letter authorizes commercial marketing of the drug with specific prescribing information for specific indications. A Complete Response Letter indicates that the review cycle of the application is complete and the application will not be approved in its present form. A Complete Response Letter usually describes all of the specific deficiencies in the BLA identified by the FDA.

The Complete Response Letter may require additional clinical or other data, additional pivotal Phase 3 clinical trial(s) and/or other significant and time-consuming requirements related to clinical trials, pre-clinical studies or manufacturing. If a Complete Response Letter is issued, the applicant may either resubmit the BLA, addressing all of the deficiencies identified in the letter, or withdraw the application. Even if such data and information are submitted, the FDA may decide that the re-submitted BLA does not satisfy the criteria for approval.

If a product receives regulatory approval, the approval is limited to the conditions of use (e.g., patient population, indication) described in the application. Further, depending on the specific risk(s) to be addressed, the FDA may require that contraindications, warnings or precautions be included in the product labeling, require that post-approval trials, including Phase 4 clinical trials, be conducted to further assess a product's safety after approval, require testing and surveillance programs to monitor the product after commercialization, or impose other conditions, including distribution and use restrictions or other risk management mechanisms under a REMS, which can materially affect the potential market and profitability of the product. The FDA may prevent or limit further marketing of a product based on the results of post-marketing trials or surveillance programs. After approval, some types of changes to the approved product, such as adding new indications, manufacturing changes and additional labeling claims, are subject to further testing requirements and FDA review and approval.

Orphan Drug Designation

Under the Orphan Drug Act, the FDA may grant orphan designation to a drug or biologic product intended to treat a rare disease or condition, which is generally a disease or condition that affects fewer than 200,000 individuals in the United States, or more than 200,000 individuals in the United States and for which there is no reasonable expectation that the cost of developing and making the product available in the United States for this type of disease or condition will be recovered from sales of the product in the United States. Orphan drug designation must be requested before submitting a BLA. After the FDA grants orphan drug designation, the identity of the therapeutic agent and its potential orphan use are disclosed publicly by the FDA. Orphan drug designation does not convey any advantage in, or shorten the duration of, the regulatory review and approval process. Orphan drug designation entitles a party to financial incentives such as opportunities for grant funding towards clinical trial costs, tax advantages and user-fee waivers. If a product that has orphan designation subsequently receives the first FDA approval for the disease or condition for which it has such designation, the product is entitled to orphan drug exclusivity, which means that the FDA may not approve any other applications to market the same drug for the same indication for seven years from the date of such approval, except in limited circumstances, such as a showing of clinical superiority to the product with orphan exclusivity by means of greater effectiveness, greater safety, by providing a major contribution to patient care or in instances of drug supply issues. Competitors, however, may receive approval of either a different product for the same indication or the same product for a different indication that could be used "off-label" by physicians in the orphan indication, even though the competitor's product is not approved in the orphan indication. Orphan drug exclusivity also could block the approval of one of our products for seven years if a competitor obtains approval before we do of the same product, as defined by the FDA, for the same indication we are seeking, or if our product candidate is determined to be contained within the scope of the competitor's product for the same indication or disease. If one of our products designated as an orphan drug receives marketing approval for an indication broader than that which is designated, it may not be entitled to orphan drug exclusivity. Orphan drug status in the European Union has similar, but not identical, requirements and benefits.

Expedited Review and Approval

The FDA has various programs, including fast track designation, breakthrough therapy designation, accelerated approval and priority review, which are intended to expedite or simplify the process for the development and FDA review of drugs and biologics that are intended for the treatment of serious or life-threatening diseases or conditions and demonstrate the potential to address unmet medical needs. The purpose of these programs is to provide important new drugs and biologics to patients earlier than under standard FDA review procedures.

To be eligible for a fast track designation, the FDA must determine, based on the request of a sponsor, that a product is intended to treat a serious or life-threatening disease or condition and demonstrates the potential to address an unmet medical need by providing a therapy where none exists or a therapy that may be potentially superior to existing therapy based on efficacy or safety factors. Fast track designation provides opportunities for more frequent interactions with the FDA review team to expedite development and review of the product. The FDA may also review sections of the BLA for a fast track product on a rolling basis before the complete application is submitted, if the sponsor and the FDA agree on a schedule for the submission of the application sections, and the sponsor pays any required user fees upon submission of the first section of the BLA.

In addition, under the provisions of the or FDASIA, passed in July 2012, a sponsor can request designation of a product candidate as a "breakthrough therapy." A breakthrough therapy is defined as a drug or biologic that is intended, alone or in combination with one or more other drugs or biologics, to treat a serious or life-threatening disease or condition, and preliminary clinical evidence indicates that the drug or biologic may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. Drugs or biologics designated as breakthrough therapies are also eligible for accelerated approval. The FDA must take certain actions with respect to breakthrough therapies, such as holding timely meetings with and providing advice to the product sponsor, intended to expedite the development and review of an application for approval of a breakthrough therapy.

Products studied for their safety and effectiveness in treating serious or life-threatening illnesses and that provide meaningful therapeutic benefit over existing treatments may be eligible for accelerated approval and may be approved on the basis of adequate and well-controlled clinical trials establishing that the product has an effect on a surrogate endpoint that is reasonably likely to predict clinical benefit, or on a clinical endpoint that can be measured earlier than IMM, that is reasonably likely to predict an effect on IMM or other clinical benefit, taking into account the severity, rarity or prevalence of the condition and the availability or lack of alternative treatments. As a condition of approval, the FDA may require a sponsor of a product receiving accelerated approval to perform post-marketing trials to verify and describe the predicted effect on IMM or other clinical endpoint, and the product may be subject to expedited withdrawal procedures.

Once a BLA is submitted for a product intended to treat a serious condition, the FDA may assign a priority review designation if the FDA determines that the product, if approved, would provide a significant improvement in safety or effectiveness. Under priority review, the FDA must review an application in six months, compared to ten months for a standard review. Most products that are eligible for fast track or breakthrough therapy designation are also likely to be considered appropriate to receive a priority review.

Even if a product qualifies for one or more of these programs, the FDA may later decide that the product no longer meets the conditions for qualification or decide that the time period for FDA review or approval will not be shortened. Furthermore, fast track designation, breakthrough therapy designation, accelerated approval and priority review do not change the standards for approval and may not ultimately expedite the development or approval process.

Post-approval Requirements

Following approval of a new product, the manufacturer and the approved product are subject to pervasive and continuing regulation by the FDA, including, among other things, monitoring and recordkeeping activities, reporting of adverse experiences with the product, product sampling and distribution restrictions, complying with promotion and advertising requirements, which include restrictions on promoting drugs for unapproved uses or patient populations (i.e., "off-label use") and limitations on industry-sponsored scientific and educational activities. Although physicians may prescribe legally available products for off-label uses, manufacturers may not market or promote such uses. The FDA and other agencies actively enforce the laws and regulations prohibiting the promotion of off-label uses, and a company that is found to have improperly promoted off-label uses may be subject to significant liability. If there are any modifications to the product, including changes in indications, labeling or manufacturing processes or facilities, the applicant may be required to submit and obtain FDA approval of a new BLA or BLA supplement, which may require the applicant to develop additional data or conduct additional pre-clinical studies and clinical trials. The FDA may also place other conditions on approvals including the requirement for a REMS to assure the safe use of the product. A REMS could include medication guides, physician communication plans or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools. Any of these limitations on approval or marketing could restrict the commercial promotion, distribution, prescription or dispensing of products. Product approvals may be withdrawn for non-compliance with regulatory standards or if problems occur following initial marketing.

FDA regulations require that products be manufactured in specific approved facilities and in accordance with cGMPs. We rely, and expect to continue to rely, on third parties for the production of clinical and commercial quantities of our products in accordance with cGMP regulations. These manufacturers must comply with cGMP regulations that require, among other things, quality control and quality assurance, the maintenance of records and documentation and the obligation to investigate and correct any deviations from cGMP. Manufacturers and other entities involved in the manufacture and distribution of approved drugs or biologics are required to register their establishments with the FDA and certain state agencies, and are subject to periodic unannounced inspections by the FDA and certain state agencies for compliance with cGMP and other laws. Accordingly, manufacturers must continue to expend time, money and effort in the area of production and quality control to maintain cGMP compliance. The discovery of conditions that violate these rules, including failure to conform to cGMPs, could result in enforcement actions, and the discovery of problems with a product after approval may result in restrictions on a product, manufacturer or holder of an approved BLA, including voluntary recall.

Once an approval or clearance of a drug is granted, the FDA may withdraw the approval if compliance with regulatory requirements and standards is not maintained or if problems occur after the product reaches the market. Later discovery of previously unknown problems with a product, including adverse events of unanticipated severity or frequency, or with manufacturing processes, or failure to comply with regulatory requirements, may result in mandatory revisions to the approved labeling to add new safety information; imposition of post-market or clinical trials to assess new safety risks; or imposition of distribution or other restrictions under a REMS program. Other potential consequences include, among other things:

- restrictions on the marketing or manufacturing of the product, complete withdrawal of the product from the market or product recalls;
- fines, warning letters or other enforcement-related letters or clinical holds on post-approval clinical trials;

- refusal of the FDA to approve pending BLAs or supplements to approved BLAs, or suspension or revocation of product approvals;
- product seizure or detention, or refusal to permit the import or export of products;
- injunctions or the imposition of civil or criminal penalties; and
- consent decrees, corporate integrity agreements, debarment, or exclusion from federal healthcare programs; or mandated modification of promotional materials and labeling and the issuance of corrective information.

Biosimilars and Exclusivity

An abbreviated approval pathway for biological products that are biosimilar to or interchangeable with an FDA-licensed reference biological product was created by the Biologics Price Competition and Innovation Act of 2009, or BPCIA, as part of the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act, or the Affordable Care Act. This amendment to the PHSA, in part, attempts to minimize duplicative testing. To date, the FDA has approved a number of biosimilars, and numerous biosimilars have been approved in Europe. The FDA has issued several guidance documents outlining its approach to reviewing and approving biosimilars.

Biosimilarity, which requires that the biological product be highly similar to the reference product notwithstanding minor differences in clinically inactive components and that there be no clinically meaningful differences between the product and the reference product in terms of safety, purity and potency, must be shown through analytical studies, animal studies and a clinical trial or trials. Interchangeability requires that a biological product be biosimilar to the reference product and that the product can be expected to produce the same clinical results as the reference product in any given patient and, for products administered multiple times to an individual, that the product and the reference product may be alternated or switched after one has been previously administered without increasing safety risks or risks of diminished efficacy relative to exclusive use of the reference biological product without such alternation or switch. Complexities associated with the larger, and often more complex, structure of biological products as compared to small molecule drugs, as well as the processes by which such products are manufactured, pose significant hurdles to implementation that are still being worked out by the FDA.

A reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, and the FDA will not accept an application for a biosimilar or interchangeable product based on the reference biological product until four years after the date of first licensure of the reference product. "First licensure" typically means the initial date the particular product at issue was licensed in the United States. Date of first licensure does not include the date of licensure of (and a new period of exclusivity is not available for) a supplement for the reference product for a subsequent application filed by the same sponsor or manufacturer of the reference product (or licensor, predecessor in interest or other related entity) for a change (not including a modification to the structure of the biological product) that results in a new indication, route of administration, dosing schedule, dosage form, delivery system, delivery device or strength or for a modification to the structure of the biological product that does not result in a change in safety, purity or potency. Therefore, one must determine whether a new product includes a modification to the structure of a previously licensed product that results in a change in safety, purity or potency to assess whether the licensure of the new product is a first licensure that triggers its own period of exclusivity. Whether a subsequent application, if approved, warrants exclusivity as the "first licensure" of a biological product is determined on a case-by-case basis with data submitted by the sponsor.

The BPCIA is complex and only beginning to be interpreted and implemented by the FDA. In addition, recent government proposals have sought to reduce the 12-year reference product exclusivity period. Other aspects of the BPCIA, some of which may impact the BPCIA exclusivity provisions, have also been the subject of recent litigation. As a result, the ultimate impact, implementation and meaning of the BPCIA is subject to significant uncertainty.

U.S. Patent Term Restoration

Depending upon the timing, duration and conditions of FDA marketing approval of our product candidates, one or more of our U.S. patents may be eligible for limited patent term extension under the Drug Price Competition and Patent Term Restoration Act of 1984, referred to as the Hatch-Waxman Amendments, and similar legislation in the European Union. The Hatch-Waxman Amendments permit a patent term extension of up to five years for a patent covering an approved product as compensation for effective patent term lost during product development and the FDA regulatory review process. However, we may not receive an extension if we fail to apply within applicable deadlines, fail to apply prior to expiration of relevant patents or otherwise fail to satisfy applicable requirements. Moreover, the length of the extension could be less than we request. Only one patent per approved product can be extended, the extension must be based on the first approval for the product, and the extension cannot extend the total patent term beyond fourteen years from approval. If we are unable to obtain patent term extension or the term of any such extension is less than we request, the period during which we can enforce our patent rights for that product will be shortened and our competitors may obtain approval to market competing products sooner.

European Union Drug Development, Review and Approval

In the European Union, our product candidates also may be subject to extensive regulatory requirements. As in the United States, medicinal products can be marketed only if a marketing authorization from the competent regulatory agencies has been obtained.

Similar to the United States, the various phases of pre-clinical and clinical research in the European Union are subject to significant regulatory controls.

The Clinical Trials Directive 2001/20/EC, the Directive 2005/28/EC on GCP, and the related national implementing provisions of the individual EU Member States govern the system for the approval of clinical trials in the European Union. Under this system, an applicant must obtain prior approval from the competent national authority of the EU Member States in which the clinical trial is to be conducted. Furthermore, the applicant may only start a clinical trial at a specific study site after the competent ethics committee has issued a favorable opinion. The clinical trial application must be accompanied by, among other documents, an IMPD (the Common Technical Document) with supporting information prescribed by Directive 2001/20/EC, Directive 2005/28/EC, where relevant the implementing national provisions of the individual EU Member States and further detailed in applicable guidance documents. All suspected unexpected serious adverse reactions to the investigated drug that occur during the clinical trial have to be reported to the competent national authority and the Ethics Committee of the Member State where they occurred.

In April 2014, the new Clinical Trials Regulation, (EU) No 536/2014 (Clinical Trials Regulation) was adopted. The Regulation is anticipated to come into application in 2019. The Clinical Trials Regulation will be directly applicable in all the EU Member States, repealing the current Clinical Trials Directive 2001/20/EC. Conduct of all clinical trials performed in the European Union will continue to be bound by currently applicable provisions until the new Clinical Trials Regulation becomes applicable. The extent to which ongoing clinical trials will be governed by the Clinical Trials Regulation will depend on when the Clinical Trials Regulation becomes applicable and on the

duration of the individual clinical trial. If a clinical trial continues for more than three years from the day on which the Clinical Trials Regulation becomes applicable the Clinical Trials Regulation will at that time begin to apply to the clinical trial.

The new Clinical Trials Regulation aims to simplify and streamline the approval of clinical trials in the European Union. The main characteristics of the regulation include: a streamlined application procedure via a single entry point, the "EU portal"; a single set of documents to be prepared and submitted for the application as well as simplified reporting procedures for clinical trial sponsors; and a harmonized procedure for the assessment of applications for clinical trials, which is divided in two parts. Part I is assessed by the competent authorities of all EU Member States in which an application for authorization of a clinical trial has been submitted (Member States concerned). Part II is assessed separately by each Member State concerned. Strict deadlines have been established for the assessment of clinical trial applications. The role of the relevant ethics committees in the assessment procedure will continue to be governed by the national law of the concerned EU Member State. However, overall related timelines will be defined by the Clinical Trials Regulation.

To obtain a marketing authorization of a drug in the European Union, we may submit marketing authorization applications, or MAA, either under the so-called centralized or national authorization procedures.

Centralized Procedure

The centralized procedure provides for the grant of a single marketing authorization following a favorable opinion by the European Medicines Agency, or EMA, that is valid in all EU member states, as well as Iceland, Liechtenstein and Norway. The centralized procedure is compulsory for medicines produced by specified biotechnological processes, products designated as orphan medicinal products, advanced-therapy medicines (such as gene-therapy, somatic cell-therapy or tissue-engineered medicines) and products with a new active substance indicated for the treatment of specified diseases, such as HIV/AIDS, cancer, diabetes, neurodegenerative disorders or autoimmune diseases and other immune dysfunctions and viral diseases. The centralized procedure is optional for products that represent a significant therapeutic, scientific or technical innovation, or whose authorization would be in the interest of public health. Under the centralized procedure the maximum timeframe for the evaluation of an MAA by the EMA is 210 days, excluding clock stops, when additional written or oral information is to be provided by the applicant in response to questions asked by the Committee of Medicinal Products for Human Use, or the CHMP. Accelerated assessment might be granted by the CHMP in exceptional cases, when a medicinal product is expected to be of a major public health interest, particularly from the point of view of therapeutic innovation. The timeframe for the evaluation of an MAA under the accelerated assessment procedure is of 150 days, excluding stop-clocks.

National Authorization Procedures

There are also two other possible routes to authorize medicinal products in several EU countries, which are available for investigational medicinal products that fall outside the scope of the centralized procedure:

- Decentralized procedure. Using the decentralized procedure, an applicant may apply for simultaneous authorization in more than one EU country of medicinal products that have not yet been authorized in any EU country and that do not fall within the mandatory scope of the centralized procedure.

- Mutual recognition procedure. In the mutual recognition procedure, a medicine is first authorized in one EU Member State, in accordance with the national procedures of that

country. Following this, further marketing authorizations can be sought from other EU countries in a procedure whereby the countries concerned agree to recognize the validity of the original, national marketing authorization.

Under the above described procedures, before granting the MA, the EMA or the competent authorities of the Member States of the EEA make an assessment of the risk-benefit balance of the product on the basis of scientific criteria concerning its quality, safety and efficacy.

European Union Regulatory Exclusivity

In the European Union, new products authorized for marketing (i.e., reference products) qualify for eight years of data exclusivity and an additional two years of market exclusivity upon marketing authorization. The data exclusivity period prevents generic applicants from relying on the pre-clinical and clinical trial data contained in the dossier of the reference product when applying for a generic marketing authorization in the European Union during a period of eight years from the date on which the reference product was first authorized in the European Union. The market exclusivity period prevents a successful generic applicant from commercializing its product in the EU until ten years have elapsed from the initial authorization of the reference product in the EU. The ten-year market exclusivity period can be extended to a maximum of eleven years if, during the first eight years of those ten years, the marketing authorization holder obtains an authorization for one or more new therapeutic indications which, during the scientific evaluation prior to their authorization, are held to bring a significant clinical benefit in comparison with existing therapies.

European Union Orphan Designation and Exclusivity

The criteria for designating an orphan medicinal product in the European Union, are similar in principle to those in the United States. Under Article 3 of Regulation (EC) 141/2000, a medicinal product may be designated as orphan if (1) it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition; (2) either (a) such condition affects no more than five in 10,000 persons in the European Union when the application is made, or (b) the product, without the benefits derived from orphan status, would not generate sufficient return in the European Union to justify investment; and (3) there exists no satisfactory method of diagnosis, prevention or treatment of such condition authorized for marketing in the European Union, or if such a method exists, the product will be of significant benefit to those affected by the condition, as defined in Regulation (EC) 847/2000. Orphan medicinal products are eligible for financial incentives such as reduction of fees or fee waivers and are, upon grant of a marketing authorization, entitled to ten years of market exclusivity for the approved therapeutic indication. The application for orphan designation must be submitted before the application for marketing authorization. The applicant will receive a fee reduction for the marketing authorization application if the orphan designation has been granted, but not if the designation is still pending at the time the marketing authorization is submitted. Orphan designation does not convey any advantage in, or shorten the duration of, the regulatory review and approval process.

The ten-year market exclusivity in the European Union may be reduced to six years if, at the end of the fifth year, it is established that the product no longer meets the criteria for orphan designation, for example, if the product is sufficiently profitable not to justify maintenance of market exclusivity. Additionally, marketing authorization may be granted to a similar product for the same indication at any time if:

- the second applicant can establish that its product, although similar, is safer, more effective or otherwise clinically superior;
- the applicant consents to a second orphan medicinal product application; or

- the applicant cannot supply enough orphan medicinal product.

Rest of the World Regulation

For other countries outside of the European Union and the United States, such as countries in Eastern Europe, Latin America or Asia, the requirements governing the conduct of clinical trials, product licensing, pricing and reimbursement vary from jurisdiction to jurisdiction. Additionally, the clinical trials must be conducted in accordance with cGCP requirements and the applicable regulatory requirements and the ethical principles that have their origin in the Declaration of Helsinki.

If we fail to comply with applicable foreign regulatory requirements, we may be subject to, among other things, fines, suspension or withdrawal of regulatory approvals, product recalls, seizure of products, operating restrictions and criminal prosecution.

Other Healthcare Laws

In addition to FDA restrictions on the marketing of pharmaceutical products, other U.S., federal and state healthcare regulatory laws restrict business practices in the pharmaceutical industry. These laws include, but are not limited to, federal and state anti-kickback, false claims, data privacy and security and physician payment and drug pricing transparency laws.

The U.S. federal Anti-Kickback Statute prohibits, among other things, any person or entity from knowingly and willfully offering, paying, soliciting, receiving or providing any remuneration, directly or indirectly, overtly or covertly, to induce or in return for purchasing, leasing, ordering or arranging for or recommending the purchase, lease or order of any good, facility, item or service reimbursable, in whole or in part, under Medicare, Medicaid or other federal healthcare programs. The term "remuneration" has been broadly interpreted to include anything of value. The Anti-Kickback Statute has been interpreted to apply to arrangements between pharmaceutical and medical device manufacturers on the one hand and prescribers, purchasers, formulary managers and beneficiaries on the other hand. Although there are a number of statutory exceptions and regulatory safe harbors protecting some common activities from prosecution, the exceptions and safe harbors are drawn narrowly. Practices that involve remuneration that may be alleged to be intended to induce prescribing, purchases or recommendations may be subject to scrutiny if they do not meet the requirements of a statutory or regulatory exception or safe harbor. Failure to meet all of the requirements of a particular applicable statutory exception or regulatory safe harbor does not make the conduct per se illegal under the U.S. federal Anti-Kickback Statute. Instead, the legality of the arrangement will be evaluated on a case-by-case basis based on a cumulative review of all its facts and circumstances. Several courts have interpreted the statute's intent requirement to mean that if any one purpose of an arrangement involving remuneration is to induce referrals of federal healthcare covered business, the statute has been violated. In addition, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation. Moreover, a claim including items or services resulting from a violation of the U.S. federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the federal civil False Claims Act. The majority of states also have anti-kickback laws, which establish similar prohibitions and in some cases may apply to items or services reimbursed by any third-party payor, including commercial insurers, or to self-pay patients.

The federal false claims and civil monetary penalties laws, including the civil False Claims Act, prohibit, among other things, any person or entity from knowingly presenting, or causing to be presented, a false, fictitious or fraudulent claim for payment to, or approval by, the federal government, knowingly making, using or causing to be made or used a false record or statement material to a false or fraudulent claim to the federal government, or knowingly making a false

statement to avoid, decrease or conceal an obligation to pay money to the U.S. federal government. A claim includes "any request or demand" for money or property presented to the U.S. government. Actions under the civil False Claims Act may be brought by the Attorney General or as a *qui tam* action by a private individual in the name of the government. Violations of the civil False Claims Act can result in very significant monetary penalties and treble damages. Several biopharmaceutical, medical device and other healthcare companies have been prosecuted under these laws for, among other things, allegedly providing free product to customers with the expectation that the customers would bill federal programs for the product. Other companies have been prosecuted for causing false claims to be submitted because of the companies' marketing of products for unapproved (e.g., or off-label), and thus non-covered, uses. In addition, the civil monetary penalties statute imposes penalties against any person who is determined to have presented or caused to be presented a claim to a federal health program that the person knows or should know is for an item or service that was not provided as claimed or is false or fraudulent. Many states also have similar fraud and abuse statutes or regulations that apply to items and services reimbursed under Medicaid and other state programs, or, in several states, apply regardless of the payor.

Violations of fraud and abuse laws, including federal and state anti-kickback and false claims laws, may be punishable by criminal and civil sanctions, including fines and civil monetary penalties, the possibility of exclusion from federal healthcare programs (including Medicare and Medicaid), disgorgement of profits and corporate integrity agreements, which impose, among other things, rigorous operational and monitoring requirements on companies. Similar sanctions and penalties, as well as imprisonment, also can be imposed upon executive officers and employees of such companies. Given the significant size of actual and potential settlements, it is expected that the government authorities will continue to devote substantial resources to investigating healthcare providers' and manufacturers' compliance with applicable fraud and abuse laws.

The federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, created additional federal criminal statutes that prohibit, among other actions, knowingly and willfully executing, or attempting to execute, a scheme to defraud any healthcare benefit program, including private third-party payors, knowingly and willfully embezzling or stealing from a healthcare benefit program, willfully obstructing a criminal investigation of a healthcare offense and knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false, fictitious or fraudulent statement in connection with the delivery of or payment for healthcare benefits, items or services. Similar to the U.S. federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation.

In addition, there has been a recent trend of increased federal and state regulation of payments made to physicians and certain other healthcare providers. The Affordable Care Act imposed, among other things, new annual reporting requirements through the Physician Payments Sunshine Act for covered manufacturers for certain payments and "transfers of value" provided to physicians and teaching hospitals, as well as ownership and investment interests held by physicians and their immediate family members. Failure to submit timely, accurately and completely the required information for all payments, transfers of value and ownership or investment interests may result in civil monetary penalties. Covered manufacturers must submit reports by the 90th day of each subsequent calendar year and the reported information is publicly made available on a searchable website. In addition, certain states require implementation of compliance programs and compliance with the pharmaceutical industry's voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government, impose restrictions on marketing practices and/or require the tracking and reporting of marketing expenditures and pricing

information as well as gifts, compensation and other remuneration or items of value provided to physicians and other healthcare professionals and entities.

We may also be subject to data privacy and security regulation by both the federal government and the states in which we conduct our business. HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act, or HITECH, and their respective implementing regulations, including the Final HIPAA Omnibus Rule published on January 25, 2013, impose specified requirements relating to the privacy, security and transmission of individually identifiable health information held by covered entities and their business associates. Among other things, HITECH made HIPAAs security standards directly applicable to "business associates," defined as independent contractors or agents of covered entities that create, receive, maintain or transmit protected health information in connection with providing a service for or on behalf of a covered entity, although it is unclear that we would be considered a "business associate" in the normal course of our business. HITECH also increased the civil and criminal penalties that may be imposed against covered entities, business associates and possibly other persons, and gave state attorneys general new authority to file civil actions for damages or injunctions in federal courts to enforce the federal HIPAA laws and seek attorney's fees and costs associated with pursuing federal civil actions. In addition, state laws govern the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and may not have the same requirements, thus complicating compliance efforts.

Similar foreign laws and regulations, which may include, for instance, applicable post-marketing requirements, anti-fraud and abuse laws and implementation of corporate compliance programs and reporting of payments or other transfers of value to healthcare professionals, may apply to us to the extent that any of our product candidates, once approved, are sold in a country other than the United States.

Because of the breadth of these laws and the narrowness of their exceptions and safe harbors, it is possible that business activities can be subject to challenge under one or more of such laws. The scope and enforcement of each of these laws is uncertain and subject to rapid change in the current environment of healthcare reform, especially in light of the lack of applicable precedent and regulations. Federal and state enforcement bodies have recently increased their scrutiny of interactions between healthcare companies and healthcare providers, which has led to a number of investigations, prosecutions, convictions and settlements in the healthcare industry.

Ensuring that business arrangements with third parties comply with applicable healthcare laws and regulations is costly and time consuming. If business operations are found to be in violation of any of the laws described above or any other applicable governmental regulations a pharmaceutical manufacturer may be subject to penalties, including civil, criminal and administrative penalties, damages, fines, disgorgement of profits, individual imprisonment, exclusion from governmental funded healthcare programs, such as Medicare and Medicaid, contractual damages, reputational harm, diminished profits and future earnings, additional reporting obligations and oversight if subject to a corporate integrity agreement or other agreement to resolve allegations of non-compliance with these laws, and curtailment or restructuring of operations, any of which could adversely affect a pharmaceutical manufacturer's ability to operate its business and the results of its operations.

Coverage, Pricing and Reimbursement

Significant uncertainty exists as to the coverage and reimbursement status of any biological products for which we obtain regulatory approval. The United States government, state legislatures and foreign governments have shown significant interest in implementing cost containment programs to limit the growth of government-paid healthcare costs, including price-controls,

restrictions on reimbursement and requirements for substitution of generic products for branded drug and biologic products. In the United States and markets in other countries, patients who are prescribed products generally rely on third-party payors to reimburse all or part of the associated healthcare costs. Providers and patients are unlikely to use our products unless coverage is provided and reimbursement is adequate to cover a significant portion of the cost of our products. If approved, sales of our product candidates will depend, in part, on the availability of coverage and adequate reimbursement from third-party payors. Third-party payors include government authorities, managed care plans, private health insurers and other organizations.

In the United States, the process for determining whether a third-party payor will provide coverage for a biological product typically is separate from the process for setting the price of such product or for establishing the reimbursement rate that the payor will pay for the product once coverage is approved. With respect to biologics, third-party payors may limit coverage to specific products on an approved list, also known as a formulary, which might not include all of the FDA-approved products for a particular indication, or place products at certain formulary levels that result in lower reimbursement levels and higher cost-sharing obligation imposed on patients. A decision by a third-party payor not to cover our product candidates could reduce physician utilization of a product. Moreover, a third-party payor's decision to provide coverage for a product does not imply that an adequate reimbursement rate will be approved. Adequate third-party reimbursement may not be available to enable a manufacturer to maintain price levels sufficient to realize an appropriate return on its investment in product development. Additionally, coverage and reimbursement for products can differ significantly from payor to payor. One third-party payor's decision to cover a particular medical product does not ensure that other payors will also provide coverage for the medical product, or will provide coverage at an adequate reimbursement rate. As a result, the coverage determination process usually requires manufacturers to provide scientific and clinical support for the use of their products to each payor separately and is a time-consuming process.

As noted above, the marketability of any products for which we receive regulatory approval for commercial sale may suffer if the government and third-party payors fail to provide coverage and adequate reimbursement. An increasing emphasis on cost containment measures in the United States has increased and we expect will continue to increase the pressure on pharmaceutical pricing. Coverage policies and third-party reimbursement rates may change at any time. Even if favorable coverage and reimbursement status is attained for one or more products for which we receive regulatory approval, less favorable coverage policies and reimbursement rates may be implemented in the future. Third-party payors are increasingly challenging the prices charged for medical products and services, examining the medical necessity and reviewing the cost-effectiveness of pharmaceutical products, in addition to questioning safety and efficacy. If third-party payors do not consider a product to be cost-effective compared to other available therapies, they may not cover that product after FDA approval or, if they do, the level of payment may not be sufficient to allow a manufacturer to sell its product at a profit.

In addition, in many foreign countries, the proposed pricing for a drug must be approved before it may be lawfully marketed. The requirements governing drug pricing and reimbursement vary widely from country to country. In the European Union, governments influence the price of products through their pricing and reimbursement rules and control of national healthcare systems that fund a large part of the cost of those products to consumers. Some jurisdictions operate positive and negative list systems under which products may only be marketed once a reimbursement price has been agreed to by the government. To obtain reimbursement or pricing approval, some of these countries may require the completion of clinical trials that compare the cost effectiveness of a particular product to currently available therapies. Other member states allow companies to fix their own prices for medicines, but monitor and control company profits. There

can be no assurance that any country that has price controls or reimbursement limitations for pharmaceutical products will allow favorable reimbursement and pricing arrangements for any of our products. The downward pressure on healthcare costs in general, particularly prescription products, has become very intense. As a result, increasingly high barriers are being erected to the entry of new products. In addition, in some countries, cross border imports from low-priced markets exert a commercial pressure on pricing within a country.

Healthcare Reform and Potential Changes to Healthcare Laws

The FDAs and other regulatory authorities' policies may change and additional government regulations may be enacted that could prevent, limit or delay regulatory approval of our product candidates. For example, in December 2016, the 21st Century Cures Act, or Cures Act, was signed into law. The Cures Act, among other things, is intended to modernize the regulation of drugs and devices and to spur innovation, but its ultimate implementation is uncertain. In addition, in August 2017, the FDA Reauthorization Act was signed into law, which reauthorized the FDAs user fee programs and included additional drug and device provisions that build on the Cures Act. If we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we are not able to maintain regulatory compliance, we may lose any marketing approval that we otherwise may have obtained and we may not achieve or sustain profitability, which would adversely affect our business, prospects, financial condition and results of operations.

A primary trend in the U.S. healthcare industry and elsewhere is cost containment. Government authorities and other third-party payors have attempted to control costs by limiting coverage and the amount of reimbursement for particular medical products and services, implementing reductions in Medicare and other healthcare funding and applying new payment methodologies. For example, in March 2010, the Affordable Care Act was enacted, which, among other things, increased the minimum Medicaid rebates owed by most manufacturers under the Medicaid Drug Rebate Program; introduced a new methodology by which rebates owed by manufacturers under the Medicaid Drug Rebate Program are calculated for drugs that are inhaled, infused, instilled, implanted or injected; extended the Medicaid Drug Rebate Program to utilization of prescriptions of individuals enrolled in Medicaid managed care plans; imposed mandatory discounts for certain Medicare Part D beneficiaries as a condition for manufacturers' outpatient drugs coverage under Medicare Part D; subjected drug manufacturers to new annual fees based on pharmaceutical companies' share of sales to federal healthcare programs; imposed a new federal excise tax on the sale of certain medical devices; created a new Patient Centered Outcomes Research Institute to oversee, identify priorities in, and conduct comparative clinical effectiveness research, along with funding for such research; created the Independent Payment Advisory Board, which, once empaneled, will have authority to recommend certain changes to the Medicare program that could result in reduced payments for prescription drugs; and established a Center for Medicare Innovation at the CMS to test innovative payment and service delivery models to lower Medicare and Medicaid spending.

Since its enactment, there have been judicial and Congressional challenges to certain aspects of the Affordable Care Act, and we expect there will be additional challenges and amendments to the Affordable Care Act in the future. The current Presidential administration and members of the U.S. Congress have indicated that they may continue to seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Most recently, the Tax Cuts and Jobs Acts was enacted, which, among other things, removes penalties for not complying with the individual mandate to carry health insurance. It is uncertain the extent to which any such changes may impact our business or financial condition.

In addition, other legislative changes have been proposed and adopted in the United States since the Affordable Care Act to reduce healthcare expenditures. These changes include the

Budget Control Act of 2011, which, among other things, included aggregate reductions of Medicare payments to providers of 2% per fiscal year and that will remain in effect through 2025 unless additional action is taken by Congress; and the American Taxpayer Relief Act of 2012, which, among other things, further reduced Medicare payments to several types of providers, including hospitals, imaging centers and cancer treatment centers and increased the statute of limitations period for the government to recover overpayments to providers from three to five years. More recently, there has been heightened governmental scrutiny over the manner in which manufacturers set prices for their marketed products, which has resulted in several Congressional inquiries and proposed bills designed to, among other things, bring more transparency to product pricing, review the relationship between pricing and manufacturer patient programs and reform government program reimbursement methodologies for pharmaceutical and biologic products.

Individual states in the United States have become increasingly active in passing legislation and implementing regulations designed to control biotechnology and pharmaceutical product pricing, including price or patient reimbursement constraints, discounts, restrictions on certain product access and marketing cost disclosure and transparency measures, and, in some cases, designed to encourage importation from other countries and bulk purchasing.

We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative or executive action, either in the United States or abroad. We expect that additional state and federal healthcare reform measures will be adopted in the future, any of which could limit the amounts that federal and state governments will pay for healthcare products and services.

Employees

As of March 31, 2018, we had 47 employees.

Facilities

Our U.S. headquarters are located in Lexington, Massachusetts, where Kiniksa US has leased approximately 25,067 square feet of office space, under a lease which expires in July 2021. We believe that our offices are sufficient to meet our current needs and that suitable additional space will be available as and when needed.

Legal Proceedings

We are not subject to any material legal proceedings.

MANAGEMENT

Executive Officers and Directors

The following table sets forth the name and position of each of our executive officers and directors and their ages as of the date of this prospectus.

Name	Age	Position
Executive Officers:		
Sanj K. Patel	48	Chief Executive Officer and Chairman of the Board
Stephen Mahoney	47	President and Chief Operating Officer
Chris Heberlig	43	Chief Financial Officer
John F. Paolini, M.D., Ph.D.	53	Chief Medical Officer
Thomas Beetham	48	Chief Legal Officer
Directors:		
Felix J. Baker, Ph.D.[2][3]	49	Director
Stephen R. Biggar, M.D., Ph.D.[3]	47	Director
Thomas R. Malley[1][3]	49	Director
Tracey L. McCain[1]	50	Director
Kimberly J. Popovits[2]	59	Director
Barry D. Quart, Pharm.D.[1][2]	61	Director

(1) Member of the audit committee.

(2) Member of the compensation committee.

(3) Member of the nominating and corporate governance committee.

Executive Officers

Sanj K. Patel has served as our Chief Executive Officer and Chairman of our board of directors since our formation in July 2015. In June 2008, Mr. Patel formed Synageva BioPharm Corp., or Synageva, a biotechnology company focused on rare diseases, where he acted as President and Chief Executive Officer until its sale to Alexion Pharmaceuticals in June 2015. Prior to Synageva, Mr. Patel held various roles at Genzyme Corporation from 1999 to 2008, most recently as head of U.S. Sales, Marketing and Commercial Operations for the Genzyme Therapeutics franchise, or Genzyme. Mr. Patel is a member of the board of directors of Syros Pharmaceuticals and BioCryst Pharmaceuticals, and, from 2013 to 2015, sat on the board of directors of Intercept Pharmaceuticals. He is also the founder and director of the Sanj K. Patel and Family Foundation, a philanthropic organization that supports charities for patients with rare and devastating diseases. Mr. Patel holds a B.Sc. with Honors from the University of the South Bank, London and completed his management and business studies at Ealing College, London and his Pharmacology research program at the Wellcome Foundation. We believe that Mr. Patel is qualified to serve on our board of directors due to his extensive business, sales and product development experience in the biotechnology industry.

Stephen Mahoney has served as our Chief Operating Officer since our formation in July 2015 and as our President since June 2017. Prior to serving as our Chief Operating Officer, Mr. Mahoney held various roles at Synageva from 2012 to 2015, most recently as Chief Commercial Officer, where he was responsible for Synageva's global commercial operations. Mr. Mahoney was also responsible for areas such as Global Sales Operations & Business Analytics, Commercial Supply Chain and Logistics, Global Procurement, Patient Services, Sales Training and Legal and Corporate Development. Prior to Synageva, Mr. Mahoney held various roles at Genzyme from 2003 to 2012, most recently as the Regional Legal Director for the Asia Pacific region, where he was responsible

for legal and healthcare compliance issues for multiple business units. Mr. Mahoney holds an M.B.A. from Boston College's Carroll School of Management, a J.D. from Boston College Law School and a B.A. from Colorado College.

Chris Heberlig has served as our Chief Financial Officer since our formation in July 2015. Prior to serving as our Chief Financial Officer, Mr. Heberlig held various roles at Synageva from 2008 to 2015, most recently serving as Senior Vice President of Finance and Business Operations. At Synageva, he led strategic tax planning, including overseeing the transfer of tax and intellectual property assets to Europe, and was responsible for global financial operations, facilities, as well as program management. Mr. Heberlig holds an M.B.A. from Boston University School of Management and a B.A. from St. Lawrence University. Mr. Heberlig is also a Certified Public Accountant.

John F. Paolini, *M.D., Ph.D.*, has served as our Chief Medical Officer since August 2016. From August 2015 to August 2016, Dr. Paolini was Clinical Research Head of the Cardiovascular and Metabolic Diseases Research Unit at Pfizer Inc., a pharmaceutical company, where he was responsible for bringing forward programs from pre-clinical through early clinical development and proof of concept. Prior to Pfizer, from August 2011 to July 2015, Dr. Paolini served as Chief Medical Officer of Cerenis Therapeutics, a biotechnology company focused on cardiovascular and metabolic diseases, where he was responsible for designing and executing clinical trials and regulatory strategy for a portfolio of products. Dr. Paolini holds an M.D., Ph.D. from Duke University School of Medicine and a B.A. and a Bachelor of Science, or B.S., from Tulane University, and completed his internship, residency and fellowship in Internal Medicine and Cardiology from Brigham and Women's Hospital, Boston.

Thomas Beetham has served as our Chief Legal Officer since our formation in July 2015 and is also responsible for corporate development. Prior to serving as our Chief Legal Officer, Mr. Beetham was the Chief Legal Officer and Senior Vice President of Corporate Development for Synageva from October 2013 to June 2015. At Synageva, in addition to leading the legal department, Mr. Beetham was responsible for business development activities. Prior to joining Synageva, from October 2011 to October 2013, Mr. Beetham was the General Legal Counsel for New England Biolabs, Inc., a reagent supplier for genomic research, where he was responsible for legal matters and a member of Biolabs' global business development team. Before Synageva, Mr. Beetham was at Genzyme from September 2004 to October 2011, most recently as the lead corporate attorney responsible for Genzyme's hematology/oncology and multiple sclerosis products, and from September 1999 to September 2004 was a corporate and transactional attorney with the law firm of Palmer & Dodge, LLP. Mr. Beetham holds an M.B.A. from Boston College's Carroll School of Management, a J.D. from Boston College Law School and a B.A. from the University of Rochester.

Directors

Felix J. Baker, Ph.D., has served as our lead director and on our board of directors since October 2015. Since 2000, Dr. Baker has been a Co-Managing Member of Baker Bros. Advisors LP, or Baker Brothers, an investment advisor focused on investments in life science and biotechnology companies. Dr. Baker and his brother, Julian Baker, started their fund management careers in 1994 when they co-founded a biotechnology investing partnership with the Tisch Family. Dr. Baker currently serves on the boards of directors of Alexion Pharmaceuticals, Genomic Health, Inc. and Seattle Genetics, Inc. and previously served on the board of directors of Synageva. Dr. Baker holds a B.S. and a Ph.D. in Immunology from Stanford University, where he also completed two years of medical school. We believe Dr. Baker is qualified to serve on our board of directors due to his extensive experience in the biotechnology industry and experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Stephen R. Biggar, M.D., Ph.D., has served as a member of our board of directors since October 2015. Since 2000, Dr. Biggar has been a partner at Baker Brothers. Dr. Biggar is currently Chairman of the board of directors of ACADIA Pharmaceuticals, serves on the board of Vivelix Pharmaceuticals, Ltd. and previously served on the board of directors of Synageva. Dr. Biggar holds an M.D. and a Ph.D. in Immunology from Stanford University and a BS in Genetics from the University of Rochester. We believe Dr. Biggar is qualified to serve on our board of directors due to his experience in the biotechnology industry, his medical and scientific training and experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Thomas R. Malley has served as a member of our board of directors since December 2016. Since May 2007, Mr. Malley has served as the President of Mossrock Capital, LLC, a private investment firm. Mr. Malley serves on the boards of directors of BeiGene, Ltd. and Kura Oncology, Inc., and previously served on the boards of directors of OvaScience, Inc., Cougar Biotechnology, Puma Biotechnology and Synageva. Mr. Malley holds a B.S. degree in Biology from Stanford University. We believe Mr. Malley is qualified to serve on our board of directors due to his experience working in the biopharmaceutical industry and experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Tracey L. McCain has served as a member of our board of directors since February 2018. Since September 2016, Ms. McCain has served as Executive Vice President and Chief Legal and Compliance Officer of Blueprint Medicine Corporation, or Blueprint, a biotechnology company. Prior to Blueprint, from January 2016 to September 2016, Ms. McCain was Senior Vice President and Head of Legal for Sanofi Genzyme, a global business unit of Sanofi,. Between joining Genzyme in May 1997 to January 2016, Ms. McCain held various roles at Genzyme, including as General Counsel following Genzyme's acquisition by Sanofi in 2011. Ms. McCain holds a J.D. from Columbia University School of Law and a B.A. from the University of Pennsylvania. We believe Ms. McCain is qualified to sit on our board of directors due to her experience working with numerous biotechnology and pharmaceutical companies.

Kimberly J. Popovits has served as a member of our board of directors since February 2018. Since 2009, Ms. Popovits has served as the Chief Executive Officer of Genomic Health, Inc., and since 2012, has served as the Chairman of the board of directors Ms. Popovits also serves on the board of directors of MyoKardia, Inc., and previously sat on the board of directors of ZS Pharma Inc. Ms. Popovits holds a B.A degree in Business from Michigan State University. We believe Ms. Popovits is qualified to sit on our board of directors due to her experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Barry D. Quart, *Pharm.D.*, has served as a member of our board of directors since October 2015. Since 2013, Dr. Quart has served as the Chief Executive Officer and on the board of directors of Heron Therapeutics, Inc., or Heron, a biotechnology company. In 2006, Dr. Quart co-founded Ardea Biosciences, Inc., a biotechnology company, and served as its President and Chief Executive Officer, and on its board of directors, from its inception through May 2013. Dr. Quart previously served on the board of directors of Synageva. Dr. Quart holds a Pharm.D. degree from the University of California, San Francisco. We believe Dr. Quart is qualified to serve on our board of directors due to his extensive management experience in the biotechnology industry and his experience developing pharmaceutical products.

Family Relationships

There are no family relationships between our board of directors and our executive officers.

Board Composition

Our board of directors is currently comprised of seven members. The members of our board of directors were elected in compliance with the provisions of the voting agreement among us and our major shareholders. The voting agreement will terminate upon the closing of this offering, and we will have no further contractual obligations regarding the election of our directors. Our directors hold office until the shareholders shall determine or, in the absence of such a determination, until the next annual general meeting or until their successors have been elected or appointed or their office is otherwise vacated.

Our amended and restated bye-laws that will become effective immediately following the closing of this offering provide that the authorized number of directors may be changed only by resolution of our board of directors. Our amended and restated bye-laws also provide that our directors may be removed only for cause by the affirmative vote of a majority of the votes entitled to be cast by our shareholders entitled to vote in an annual general election of directors. Any vacancy on our board of directors, including a vacancy resulting from an enlargement of our board of directors or from removal for cause not filled by the shareholders at the time, may be filled only by vote of a majority of our directors then in office.

In accordance with the terms of our amended and restated bye-laws that will become effective immediately following the closing of this offering, our board of directors will be divided into three classes, Class I, Class II and Class III. At the first general meeting of shareholders following this offering, each Class I director shall be elected for a three-year initial term of office, each Class II director shall be elected for a one-year initial term of office, and each Class III director shall be elected for a two-year initial term of office. Thereafter, members of each class shall serve three-year terms. Upon the closing of this offering, the members of the classes will be divided as follows:

- the Class I directors will be Sanj K. Patel and Thomas R. Malley;
- the Class II directors will be Stephen R. Biggar and Barry D. Quart; and
- the Class III directors will be Felix J. Baker, Tracey L. McCain and Kimberly J. Popovits.

Upon the expiration of the term of a class of directors, directors in that class will be eligible to be elected for a new three-year term at the annual meeting of shareholders in the year in which their term expires.

We have no formal policy regarding board diversity. Our priority in selection of board members is identification of members who will further the interests of our shareholders through his or her established record of professional accomplishment, the ability to contribute positively to the collaborative culture among board members, knowledge of our business and understanding of the competitive landscape.

In selecting board members for nomination, our board may consider many factors, such as personal and professional integrity; experience in corporate management, such as serving as an officer or former officer of a pharmaceutical or biotechnology company; experience as a board member or executive officer of another publicly-held company; diversity of expertise and experience in substantive matters pertaining to our business relative to other board members; and diversity of background and perspective, including, but not limited to, with respect to age, gender, race, place of residence and specialized experience.

Director Independence

Our board of directors has determined that, of our seven directors, Felix J. Baker, Stephen R. Biggar, Thomas R. Malley, Tracey L. McCain, Kimberly J. Popovits and Barry D. Quart do not have a relationship that would interfere with the exercise of independent judgment in carrying out the

responsibilities of a director and that each of these directors is "independent" as that term is defined under the rules of The Nasdaq Stock Market LLC. There are no family relationships among any of our directors or executive officers.

Board Leadership Structure

Our board of directors is currently chaired by our Chief Executive Officer, Sanj K. Patel, and our lead independent director is Felix J. Baker, Ph.D. Our corporate governance guidelines provide the flexibility for our board of directors to modify our leadership structure in the future as it deems appropriate.

Role of the Board in Risk Oversight

One of the key functions of our board of directors is informed oversight of our risk management process. Our board of directors does not have a standing risk management committee, but rather administers this oversight function directly through our board of directors as a whole, as well as through various standing committees of our board of directors that address risks inherent in their respective areas of oversight. In particular, our board of directors is responsible for monitoring and assessing strategic risk exposure and our audit committee has the responsibility to consider and discuss our major financial risk exposures and the steps our management has taken to monitor and control these exposures, including guidelines and policies to govern the process by which risk assessment and management is undertaken. Our audit committee also monitors compliance with legal and regulatory requirements. Our nominating and corporate governance committee monitors the effectiveness of our corporate governance practices, including whether they are successful in preventing illegal or improper liability-creating conduct. Our compensation committee assesses and monitors whether any of our compensation policies and programs has the potential to encourage excessive risk-taking. While each committee is responsible for evaluating certain risks and overseeing the management of such risks, our entire board of directors is regularly informed through committee reports about such risks.

Board Committees

Our board of directors has established three standing committees — audit, compensation and nominating and corporate governance — each of which operates under a charter that has been approved by our board of directors. Upon our listing on Nasdaq, each committee's charter will be available under the Corporate Governance section of our website at *www.kiniksa.com*. The reference to our website address does not constitute incorporation by reference of the information contained at or available through our website, and you should not consider it to be a part of this prospectus.

Audit Committee

The audit committee's responsibilities include:

- appointing, approving the compensation of, and assessing the independence of our registered public accounting firm;
- overseeing the work of our registered public accounting firm, including through the receipt and consideration of reports from such firm;
- reviewing and discussing with management and the registered public accounting firm our annual and quarterly financial statements and related disclosures;
- coordinating our board of directors' oversight of our internal control over financial reporting, disclosure controls and procedures and code of business conduct and ethics;

- discussing our risk management policies;
- meeting independently with our internal auditing staff, if any, registered public accounting firm and management;
- reviewing and approving or ratifying any related person transactions; and
- preparing the audit committee report required by SEC rules.

The members of our audit committee are Thomas R. Malley, Tracey L. McCain and Barry D. Quart. Mr. Malley serves as the chairperson of the committee. All members of our audit committee meet the requirements for financial literacy under the applicable listing rules of Nasdaq. Our board of directors has determined that Mr. Malley, Ms. McCain and Dr. Quart meet the independence requirements of Rule 10A-3 under the Exchange Act and the applicable Nasdaq rules. Our board of directors has determined that Mr. Malley is an "audit committee financial expert" as defined by applicable SEC rules and has the requisite financial sophistication as defined under the applicable Nasdaq rules.

Compensation Committee

The compensation committee's responsibilities include:

- reviewing and approving, or recommending for approval by the board of directors, the compensation of our Chief Executive Officer and our other executive officers;
- overseeing and administering our cash and equity incentive plans;
- reviewing and making recommendations to our board of directors with respect to director compensation;
- reviewing and discussing annually with management our "Compensation Discussion and Analysis" to the extent required; and
- preparing the annual compensation committee report required by SEC rules, to the extent required.

The members of our compensation committee are Felix J. Baker, Kimberly J. Popovits and Barry D. Quart. Dr. Baker serves as the chairperson of the committee. Our board of directors has determined that Drs. Baker and Quart and Ms. Popovits are independent under the applicable Nasdaq rules, including the Nasdaq rules specific to membership on the compensation committee and that each is a "non-employee director" as defined in Rule 16b-3 promulgated under the Exchange Act.

Nominating and Corporate Governance Committee

The nominating and corporate governance committee's responsibilities include:

- identifying individuals qualified to become board members;
- recommending to our board of directors the persons to be nominated for election as directors and to each board committee; and
- overseeing a periodic evaluation of our board of directors.

The members of our nominating and corporate governance committee are Felix J. Baker, Stephen R. Biggar and Thomas R. Malley. Dr. Biggar serves as the chairperson of the committee. Our board of directors has determined that Drs. Biggar and Baker and Mr. Malley are independent under the applicable Nasdaq rules.

Compensation Committee Interlocks and Insider Participation

No member of our compensation committee is or has been our current or former officer or employee. None of our executive officers served as a director or a member of a compensation committee (or other committee serving an equivalent function) of any other entity, one of whose executive officers served as a director or member of our compensation committee during the fiscal year ended December 31, 2017.

Code of Ethics and Code of Conduct

We have adopted a written code of business conduct and ethics that applies to our directors, officers and employees, including our principal executive officer, principal financial officer, principal accounting officer or controller, or persons performing similar functions. Upon our listing on Nasdaq, our code of business conduct and ethics will be available under the Corporate Governance section of our website at *www.kiniksa.com*. In addition, we intend to post on our website all disclosures that are required by law or the Nasdaq rules concerning any amendments to, or waivers from, any provision of the code. The reference to our website address does not constitute incorporation by reference of the information contained at or available through our website, and you should not consider it to be a part of this prospectus.

EXECUTIVE AND DIRECTOR COMPENSATION

This section discusses the material components of the executive compensation program for our executive officers who are named in the "2017 Summary Compensation Table" below. In 2017, our "named executive officers" and their positions were as follows:

- Sanj K. Patel, our Chief Executive Officer and Chairman of the Board of Directors;
- Stephen Mahoney, our President and Chief Operating Officer; and
- John F. Paolini, M.D., Ph.D., our Chief Medical Officer.

This discussion may contain forward-looking statements that are based on our current plans, considerations, expectations and determinations regarding future compensation programs. Actual compensation programs that we adopt following the completion of this offering may differ materially from the currently planned programs summarized in this discussion.

2017 Summary Compensation Table

The following table sets forth information concerning the compensation of our named executive officers for the year ended December 31, 2017.

Name and Principal Position	Year	Salary ($)	Option Awards ($)[1]	Non-Equity Incentive Plan Compensation ($)	All Other Compensation ($)[2]	Total ($)
Sanj K. Patel *Chief Executive Officer and Chairman of the Board*	2017	700,000	644,950	140,000	10,800	1,495,750
Stephen Mahoney *President and Chief Operating Officer*	2017	405,000	219,629	81,000	10,800	716,429
John F. Paolini, M.D., Ph.D. *Chief Medical Officer*	2017	380,000	275,358	114,000	10,800	780,158

(1) Amounts reflect the full grant-date fair value of share options granted during 2017 computed in accordance with ASC Topic 718, rather than the amounts paid to or realized by the named individual. We provide information regarding the assumptions used to calculate the value of all option awards in Note 8 to our consolidated financial statements included in this prospectus.

(2) Amount shown represents 401(k) matching contributions. For additional information, refer to the discussion in the "Narrative Disclosure to Summary Compensation Table" below under the heading "— Other Elements of Compensation — Retirement Plans."

Narrative Disclosure to Summary Compensation Table

The primary elements of compensation for our named executive officers are base salary, annual performance bonuses and long-term equity-based compensation awards. The named executive officers also generally participate in employee benefit plans and programs that we offer to our other full-time employees on the same basis.

2017 Salaries

We pay our named executive officers a base salary that is intended to provide a fixed component of compensation reflecting the executive's skill set, experience, role and responsibilities. Base salaries for our named executive officers generally have been set at levels deemed necessary to attract and retain the named executive officers and were originally established in each named

executive officer's employment agreement or offer letter. The following table shows the annual base salaries for 2017 of our named executive officers:

Name	2017 Annual Base Salary ($)
Sanj K. Patel	700,000
Stephen Mahoney	405,000
John F. Paolini, M.D., Ph.D.	380,000

2017 Bonuses

We offer our named executive officers the opportunity to earn annual performance bonuses to compensate them for attaining short-term company and individual goals as approved by our board of directors. For 2017, performance bonuses were based on attaining corporate goals relating to the overall business, including development of KPL-716, business development, intellectual property protection, supply chain requirements, key employee retention and recruitment, capitalization and cost and expense control and operational compliance. The 2017 target bonus amounts for our named executive officers, expressed as percentages of their respective annual base salaries, were 10% for Mr. Patel, 10% for Mr. Mahoney and 30% for Dr. Paolini.

In December 2017, our board of directors met to review performance against the 2017 bonus goals and approved cash bonuses for the named executive officers in the amounts set forth in the Non-Equity Incentive Plan Compensation column of the 2017 Summary Compensation Table above.

Equity Compensation

We generally offer share options to our employees, including our named executive officers, as the long-term incentive component of our compensation program. Share options allow our employees to purchase our Class A common shares at a price equal to the fair market value per Class A common share on the date of grant, as determined by the board of directors. In 2017, our named executive officers were granted the share options set forth in the table below. The options vest over four years from the applicable grant date with 25% of the option vesting on the first anniversary of the grant date and 2.0833% of the shares vesting monthly for three years thereafter.

Named Executive Officer	2017 Option Awards Granted
Sanj K. Patel	257,969
Stephen Mahoney	87,848
John F. Paolini, M.D., Ph.D.	110,138

These options were issued under our 2015 Equity Incentive Plan, or the 2015 Plan, with exercise prices equal to the fair market value of our Class A common shares on the date of grant, as determined by the board of directors, and subject to our standard vesting schedule described above.

In connection with this offering, our board of directors adopted, and our shareholders have approved, the 2018 Incentive Award Plan, or the 2018 Plan, in order to facilitate the grant of cash and equity incentives to directors, employees (including our named executive officers) and consultants and to enable us to obtain and retain services of these individuals, which we believe is essential to our long-term success. The 2018 Plan will be effective on the day prior to the first public trading date of our Class A common shares. Following the effective date of the 2018 Plan, we will not make any further grants under our 2015 Plan. However, the 2015 Plan will continue to govern the terms and conditions of the outstanding awards granted under it. For additional

information about the 2015 Plan and the 2018 Plan, please see the section titled "Executive Compensation Plans" below.

Other Elements of Compensation

Retirement Plans

We maintain a 401(k) retirement savings plan for our employees, including our named executive officers, who satisfy certain eligibility requirements. Our named executive officers are eligible to participate in the 401(k) plan on the same terms as other full-time employees. We provide matching contributions of 100% of the first 3% of each participant's salary contributed, plus 50% for each of the next 2% contributed. Employee contributions are allocated to each participant's individual account and are then invested in selected investment alternatives according to the participants' directions. Employees are immediately and fully vested in their own contributions and the employer match. We believe that providing a vehicle for tax-deferred retirement savings though our 401(k) plan adds to the overall desirability of our executive compensation package and further incentivizes our employees, including our named executive officers, in accordance with our compensation policies.

Employee Benefits and Perquisites

All of our full-time employees, including our named executive officers, are eligible to participate in our health and welfare plans, including medical, dental and vision benefits, a healthcare spending account, a dependent care flexible spending account, short-term and long-term disability insurance and life insurance to the same extent as our other full-time employees generally, subject to the terms and eligibility requirements of those plans.

No Tax Gross-Ups

We do not make gross-up payments to cover our named executive officers' personal income taxes that may pertain to any of the compensation or perquisites paid or provided by us.

Outstanding Equity Awards at 2017 Fiscal Year-End

The following table summarizes the number of Class A common shares underlying outstanding equity incentive plan awards for each named executive officer as of December 31, 2017.

		Option Awards			
Name	Vesting Start Date	Number of Securities Underlying Unexercised Options (#) Exercisable	Number of Securities Underlying Unexercised Options (#) Unexercisable[1]	Option Exercise Price ($)	Option Expiration Date
Sanj K. Patel	8/1/2015	293,864	209,904	1.59	12/15/2025
	6/28/2017	—	257,969	3.80	6/28/2027
Stephen Mahoney	8/1/2015	73,466	52,475	1.59	12/15/2025
	6/28/2017	—	87,848	3.80	6/28/2027
John F. Paolini, M.D., Ph.D.	8/15/2016	75,858	151,716	1.86	9/13/2026
	6/28/2017	—	110,138	3.80	6/28/2027

(1) The options vest over a four-year period with 25% of the shares vesting on the first anniversary of the corresponding vesting start date, and 2.0833% of the shares vesting monthly for three years thereafter. Pursuant to each named executive officer's employment agreement in effect on December 31, 2017, in the event of a termination of employment by the Company without Cause or a result of the named executive officer's death, disability or resignation for Good Reason (as such capitalized terms are defined in their respective employment agreements), Mr. Mahoney and Dr. Paolini are entitled to accelerated vesting of all of their then-unvested Company equity or equity-based awards that would have, absent termination, become vested within 12 months following termination, and Mr. Patel is entitled to accelerated vesting of all of his then-unvested equity or equity-based awards that would have, absent termination, become vested within 18 months following termination. In addition, in the event of a change in control (as defined in the applicable option award agreement), each named executive officer will become immediately 100% fully vested in the named executive officer's option to the extent that such award is not assumed or substituted.

Executive Compensation Arrangements

In connection with this offering, we intend to amend the existing employment agreements with each of our named executive officers. Certain key terms of these agreements are described below.

Sanj K. Patel

The term of our amended and restated employment agreement with Mr. Patel lasts until either the Company or Mr. Patel terminates his employment by giving notice to the other party or his employment terminates due to his death. Pursuant to the amended and restated employment agreement, Mr. Patel is entitled to receive an annual base salary of at least $750,000, subject to increase from time to time by the Company and the opportunity to earn an annual performance-based bonus based on actual corporate and individual performance against established objectives for each calendar year, with a minimum target bonus opportunity of 60% of his annual base salary. In addition, if Mr. Patel's employment with us is terminated as a result of his death or disability, by the Company without Cause, or by Mr. Patel for Good Reason, whether or not in connection with a change in control, he will be entitled to receive (a) a lump sum payment equal to (i) 200% of the sum of his annual base salary and the target bonus for the year of termination plus (ii) $25,000 and (b) a prorated portion of his target bonus for the year of termination. Also, if the termination occurs

other than during the 12-month period following a change in control, Mr. Patel will be entitled to accelerated vesting of all of his then-unvested time-vesting equity that would have, absent termination, become vested within 18 months following termination, or if the termination occurs during the 12-month period following a change in control, Mr. Patel will be entitled to full accelerated vesting of all of his then-unvested time-vesting equity. Mr. Patel's right to receive these severance payments and benefits is subject to his execution and non-revocation of a release of claims for the benefit of the Company and his compliance with certain confidentiality obligations and restrictive covenants.

In the event of a change in control, Mr. Patel will become immediately 100% fully vested in each time-vesting equity award granted to him that is not assumed or substituted for in the change in control transaction.

Stephen Mahoney

The term of our amended and restated employment agreement with Mr. Mahoney lasts until either the Company or Mr. Mahoney terminates his employment by giving notice to the other party or his employment terminates due to his death. Pursuant to the amended and restated employment agreement, Mr. Mahoney is entitled to receive an annual base salary of $442,260, subject to change from time to time by the Company, and the opportunity to earn an annual performance-based bonus, with an initial target bonus opportunity of 40% of his annual base salary. In addition, if Mr. Mahoney's employment with us is terminated as a result of his death or disability, by the Company without Cause, or by Mr. Mahoney for Good Reason, he will be entitled to receive (a) a lump sum payment equal to his annual base salary plus $16,500, (b) a prorated portion (or, if the termination occurs during the 12 months following a change in control, 100%) of his target bonus for the year of termination and (c) accelerated vesting of all of his then-unvested time-vesting equity that would have, absent termination, become vested within 12 months following termination (or, if the termination occurs during the 12 months following a change in control, full accelerated vesting of all of his then-unvested time-vesting equity). Mr. Mahoney's right to receive these severance payments and benefits is subject to his execution and non-revocation of a release of claims for the benefit of the Company and his compliance with certain confidentiality obligations and restrictive covenants.

John F. Paolini, M.D., Ph.D.

The term of our amended and restated employment agreement with Dr. Paolini lasts until either the Company or Dr. Paolini terminates his employment by giving notice to the other party or his employment terminates due to his death. Pursuant to the amended and restated employment agreement, Dr. Paolini is entitled to receive an annual base salary of $420,000, subject to change from time to time by the Company, and the opportunity to earn a discretionary performance-based bonus, with a target bonus opportunity of 30% of his annual base salary. In addition, if Dr. Paolini's employment with us is terminated as a result of his death or disability or without Cause, he will be entitled to receive (a) a lump sum payment that is equivalent to 9 months of his annual base salary (or, if the termination occurs during the 12 months following a change in control, a lump sum payment that is equivalent to 12 months of his annual base salary) plus $16,500, (b) a prorated portion (or, if the termination occurs during the 12 months following a change in control, 100%) of his target bonus for the year of termination and (c) accelerated vesting of all of his then-unvested time-vesting equity that would have, absent termination, become vested within 12 months following termination (or, if the termination occurs during the 12 months following a change in control, full accelerated vesting of all of his then-unvested time-vesting equity). Dr. Paolini's right to receive these severance payments and benefits is subject to his execution and non-revocation of a release

of claims for the benefit of the Company and his compliance with certain confidentiality obligations and restrictive covenants.

As used in the executive employment agreements, the following capitalized terms generally have the following meanings:

- The term Cause generally means (i) gross negligence or willful misconduct in performance of the named executive officer's duties which results in material damage to us; (ii) the commission of any act of fraud, embezzlement or professional dishonesty with respect to our business; (iii) the commission of a felony or crime involving moral turpitude; (iv) the material breach of any provision of the executive employment agreement or any other written agreement between the named executive officer and us; or (v) the failure to comply with our lawful directives, which results in damage to us.

- The term Good Reason generally means the occurrence of any of the following events without the named executive officer's written consent: (i) a demotion or, in the case of Mr. Patel only, the assignment of duties materially inconsistent with his title, position, status, reporting relationships, authority, duties or responsibilities with us; (ii) a requirement that the named executive officer relocate his primary reporting location to a location more than fifty (50) miles from our offices in Lexington, Massachusetts; (iii) our breach of the executive employment agreement with the named executive officer; (iv) our failure to comply with the provisions addressing each named executive officer's compensation and benefits, including the base salary, bonus compensation, and annual vacation, other than insubstantial or inadvertent failures not in bad faith that we remedy promptly after receiving notice thereof; (v) for Mr. Patel only, a material diminution in the budget over which he has responsibility; or (vi) for Mr. Mahoney only, a reduction of more than five percent of his base salary other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated.

Director Compensation

Directors who are also our employees or who are affiliated with one of our principal shareholders do not receive compensation for their service as directors. Our non-employee directors have historically received a cash payment of $10,000 per year, paid quarterly, and awards of our share options as compensation for their service as directors.

2017 Director Compensation Table

The following table sets forth in summary form information concerning the compensation that was earned by or paid to each of our non-employee directors during the year that ended December 31, 2017:

Name	Fees Earned or Paid in Cash ($)	Option Awards ($)[1]	Total ($)
Felix J. Baker, Ph.D.	—	—	—
Stephen R. Biggar, M.D., Ph.D.	—	—	—
Thomas R. Malley	10,000	68,625	78,625
Barry D. Quart, Pharm.D	10,000	68,625	78,625

(1) Amounts reflect the full grant-date fair value of share options granted during 2017 computed in accordance with ASC Topic 718, rather than the amounts paid to or realized by the named individual. We provide information regarding the assumptions used to calculate the value of all option awards in Note 8 to our consolidated financial statements included in this prospectus.

The following table sets forth the aggregate numbers of share options (exercisable and unexercisable) held as of December 31, 2017 by our non-employee directors as of December 31, 2017. Refer to our Outstanding Equity Awards at 2017 Fiscal Year End table for information regarding equity awards held by Mr. Patel as of December 31, 2017.

Name	Option Awards (#)
Felix J. Baker, Ph.D.	—
Stephen R. Biggar, M.D., Ph.D.	—
Thomas R. Malley	49,407
Barry D. Quart, Pharm.D	49,407

Recent Developments Regarding Director Compensation

In May 2018, in anticipation of this offering, our board of directors approved the following grants of options to purchase Class A common shares for our non-employee directors, effective on the date that the registration statement of which this prospectus forms a part becomes effective: Felix J. Baker: 18,760 shares; Stephen R. Biggar: 18,760 shares. The options will have an exercise price equal to the initial public offering price of our common shares and will generally vest in twelve equal monthly installments occurring on the individual's completion of each month of service to the Company following the effective date of grant.

In addition, effective on the effectiveness of the registration statement of which this prospectus forms a part, our board of directors adopted and, prior to commencing this offering, our shareholders approved a compensation program for our non-employee directors under which each non-employee director will receive the following amounts for their services on our board of directors:

- an option to purchase 37,965 Class A common shares upon the director's initial election or appointment to our board of directors that occurs after our initial public offering,
- if the director has served on our board of directors for at least six months as of the date of an annual meeting of shareholders, an option to purchase 18,760 Class A common shares on the date of the annual meeting,
- an annual director fee of $35,000, and
- if the director serves on a committee of our board of directors or in the other capacities stated below, an additional annual fee as follows:
    - chairman of the board or lead independent director, $22,500,
    - chairman of the audit committee, $15,000,
    - audit committee member other than the chairman, $7,500,
    - chairman of the compensation committee, $10,000,
    - compensation committee member other than the chairman, $5,000,
    - chairman of the nominating and corporate governance committee, $8,000, and
    - nominating and corporate governance committee member other than the chairman, $4,000.

Share options granted to our non-employee directors under the program will have an exercise price equal to the fair market value of our Class A common shares on the date of grant and will expire not later than ten years after the date of grant. The share options granted upon a director's

initial election or appointment will vest and become exercisable as to one-third of the shares on the first anniversary of the date of grant and as to the remainder in twenty-four substantially equal monthly installments thereafter, subject to the director continuing in service through each such vesting date. The share options granted annually to directors will vest and become effective in twelve substantially equal monthly installments following the date of grant, subject to the director continuing in service through each such vesting date. In addition, all unvested share options will vest in full upon the occurrence of a change in control.

Director fees under the program will be payable in arrears in four equal quarterly installments not later than the fifteenth day following the final day of each calendar quarter, provided that the amount of each payment will be prorated for any portion of a quarter that a director is not serving on our board and no fee will be payable in respect of any period prior to the effective date of the registration statement of which this prospectus is a part.

Executive Compensation Plans

The following summarizes the material terms of the long-term incentive compensation plans in which our named executive officers and directors will be eligible to participate following the consummation of this offering, and the 2015 Plan, under which we have previously made periodic grants of equity and equity-based awards to our named executive officers and directors.

2015 Equity Incentive Plan

Our board of directors and shareholders have approved the 2015 Plan, under which we may grant share options, share grants and share-based awards to employees, directors and consultants. We have reserved a total of 4,794,266 of our Class A common shares for issuance under the 2015 Plan. Following the effectiveness of the 2018 Plan, we will not make any further grants under the 2015 Plan and as of the date of this prospectus, only share options are outstanding under the plan. However, the 2015 Plan will continue to govern the terms and conditions of the outstanding awards granted under it. Our Class A common shares subject to awards granted under the 2015 Plan that are forfeited, lapse unexercised or are settled in cash following the effective date of the 2018 Plan will be available for issuance under the 2018 Plan.

Administration. Our board of directors administers the 2015 Plan and has the authority to interpret the provisions of the 2015 Plan and awards outstanding thereunder, to make all rules and determinations which it deems necessary or advisable for the administration of the 2015 Plan, to determine which employees, directors and consultants will be granted awards, to determine the number of shares for which awards will be granted, to specify the terms and conditions upon which awards can be granted, to specify the terms and conditions of award agreements under the 2015 Plan, and to make all other determinations in the judgment of the board of directors that are necessary and desirable for the administration of the 2015 Plan. The board of directors may delegate its authority under the 2015 Plan to a committee. Following the effectiveness of this offering, we anticipate that the board of directors will delegate its general administrative authority under the 2015 Plan to its Compensation Committee.

Types of Awards. The 2015 Plan provides for the grant of share options, including share options intended to qualify as incentive stock options, or ISOs, under the U.S. Internal Revenue Code of 1986, as amended, or the Code, share grants and share-based awards to employees, directors and consultants of the company or its affiliates, except that share options intended to qualify as ISOs may only be granted to employees who are residents of the United States.

Certain Transactions. If certain changes are made in, or events occur with respect to, our Class A common shares, the 2015 Plan and outstanding awards will be adjusted in the class,

number and, as applicable, exercise price of securities as determined by the board of directors. In the event of certain corporate transactions of our company, including an amalgamation, consolidation, merger or sale of all or substantially all of our assets, our board or the board of directors of any corporation assuming the obligations under the 2015 Plan, may, in its discretion, take any one or more of the following actions, as to some or all options or share-based awards outstanding under the 2015 Plan (and need not take the same action as to each such option or share-based award): (i) make appropriate provisions for the continuation of options by substituting on an equitable basis for the shares then subject to options either the consideration payable with respect to the outstanding Class A common shares or securities of any successor or acquiring entity; (ii) upon written notice to the participants, provide that the options will terminate unless they are exercised within a specified number of days of the date of such notice; (iii) terminate the options in exchange for payment of an amount equal to the consideration payable upon consummation of such transaction to a holder of the number of Class A common shares into which such option would have been exercisable, less the aggregate exercise price; (iv) make appropriate provision for the continuation of such share grants on the same terms and conditions by substituting on an equitable basis for the shares then subject to the share grants either the consideration payable with respect to such outstanding shares in connection with the transaction or securities of any successor or acquiring entity; and (v) provide that, upon consummation of the transaction, each outstanding share grant shall be terminated in exchange for a payment of an amount equal to the consideration payable upon consummation of such transaction to a holder of the number of Class A common shares comprising the share grant.

Amendment and Termination. The board of directors may terminate, modify or amend the 2015 Plan from time to time, provided that any amendment or modification may not adversely affect the rights of a holder of an outstanding award without such holder's consent. The board of directors may amend or modify the 2015 Plan and any outstanding ISOs to the extent necessary to qualify any or all such options for favorable federal income tax treatment; however, any amendment approved by the board of directors which is determined to be of a scope that requires shareholder approval will be subject to obtaining such approval before taking effect.

2018 Incentive Award Plan

Our board of directors adopted and our shareholders have approved, effective the day prior to the first public trading date of our Class A common shares, the 2018 Plan, under which we may grant cash and equity-based incentive awards to eligible service providers in order to attract, retain and motivate the persons who make important contributions to our company. The material terms of the 2018 Plan are summarized below.

Eligibility and Administration

Our employees, consultants and directors, and employees and consultants of our subsidiaries, will be eligible to receive awards under the 2018 Plan. The 2018 Plan will be administered by our board of directors, which may delegate its duties and responsibilities to one or more committees of our directors and/or officers (referred to collectively as the plan administrator below), subject to the limitations imposed under the 2018 Plan, Section 16 of the Exchange Act, stock exchange rules and other applicable laws. The plan administrator will have the authority to take all actions and make all determinations under the 2018 Plan, to interpret the 2018 Plan and award agreements and to adopt, amend and repeal rules for the administration of the 2018 Plan as it deems advisable. The plan administrator will also have the authority to grant awards, determine which eligible service providers receive awards, and set the terms and conditions of all awards under the 2018 Plan, including any vesting and vesting acceleration provisions, subject to the conditions and limitations in the 2018 Plan.

Shares Available for Awards

An aggregate of 4,466,500 Class A common shares will initially be available for issuance under the 2018 Plan. The number of Class A common shares initially available for issuance will be increased by an annual increase on January 1 of each calendar year beginning in 2019 and ending in and including 2028, equal to the lesser of (A) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (B) a smaller number of our Class A common shares determined by our board of directors. No more than 27,915,000 Class A common shares may be issued under the 2018 Plan upon the exercise of ISOs. Shares issued under the 2018 Plan may be designated but unissued shares, shares purchased on the open market or treasury shares.

If an award under the 2018 Plan or the 2015 Plan, expires, lapses or is terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited, any unused shares subject to the award will, as applicable, become or again be available for new grants under the 2018 Plan. Awards granted under the 2018 Plan in substitution for any options or other share or share-based awards granted by an entity before the entity's merger or consolidation with us or our acquisition of the entity's property or shares will not reduce the shares available for grant under the 2018 Plan, but may count against the maximum number of shares that may be issued upon the exercise of ISOs.

Awards

The 2018 Plan provides for the grant of options to purchase shares, including ISOs, non-qualified share options, or NSOs, share appreciation rights, or SARs, restricted shares, dividend equivalents, restricted share units, or RSUs, and other share or cash based awards. Certain awards under the 2018 Plan may constitute or provide for payment of "nonqualified deferred compensation" under Section 409A of the Code. All awards under the 2018 Plan will be set forth in award agreements, which will detail the terms and conditions of awards, including any applicable vesting and payment terms and post-termination exercise limitations. A brief description of each award type follows.

- *Share Options and SARs.* Share options provide for the purchase of our Class A common shares in the future at an exercise price set on the grant date. ISOs, by contrast to NSOs, may provide tax deferral beyond exercise and favorable capital gains tax treatment to their holders if certain holding period and other requirements of the Code are satisfied. SARs entitle their holder, upon exercise, to receive from us an amount equal to the appreciation of the shares subject to the award between the grant date and the exercise date. The plan administrator will determine the number of shares covered by each option and SAR, the exercise price of each option and SAR and the conditions and limitations applicable to the exercise of each option and SAR. The exercise price of a share option or SAR will not be less than 100% of the fair market value of the underlying share on the grant date (or 110% in the case of ISOs granted to certain significant shareholders), except with respect to certain substitute awards granted in connection with a corporate transaction. The term of a share option or SAR may not be longer than ten years (or five years in the case of ISOs granted to certain significant shareholders).

- *Restricted Shares and RSUs.* Restricted shares are an award of nontransferable Class A common shares that remain forfeitable unless and until specified conditions are met and which may be subject to a purchase price. RSUs are contractual promises to deliver Class A common shares in the future, which may also remain forfeitable unless and until specified conditions are met and may be accompanied by the right to receive the equivalent value of dividends paid our Class A common shares prior to the delivery of the underlying shares.

The plan administrator may provide that the delivery of the shares underlying RSUs will be deferred on a mandatory basis or at the election of the participant. The terms and conditions applicable to restricted shares and RSUs will be determined by the plan administrator, subject to the conditions and limitations contained in the 2018 Plan.

- *Other Share or Cash Based Awards.* Other share or cash based awards are awards of cash, fully vested Class A common shares and other awards valued wholly or partially by referring to, or otherwise based on, our Class A common shares or other property. Other share or cash based awards may be granted to participants and may also be available as a payment form in the settlement of other awards, as standalone payments and as payment in lieu of compensation to which a participant is otherwise entitled. The plan administrator will determine the terms and conditions of other share or cash based awards, which may include any purchase price, performance goal, transfer restrictions and vesting conditions.

Certain Transactions

In connection with certain corporate transactions and events affecting our Class A common shares, including a change in control, or change in any applicable laws or accounting principles, the plan administrator has broad discretion to take action under the 2018 Plan to prevent the dilution or enlargement of intended benefits, facilitate the transaction or event or give effect to the change in applicable laws or accounting principles. This includes canceling awards for cash or property, accelerating the vesting of awards, providing for the assumption or substitution of awards by a successor entity, adjusting the number and type of shares subject to outstanding awards and/or with respect to which awards may be granted under the 2018 Plan and replacing or terminating awards under the 2018 Plan. In addition, in the event of certain non-reciprocal transactions with our shareholders, the plan administrator will make equitable adjustments to the 2018 Plan and outstanding awards as it deems appropriate to reflect the transaction.

Plan Amendment and Termination

Our board of directors may amend or terminate the 2018 Plan at any time; however, no amendment, other than an amendment that increases the number of shares available under the 2018 Plan, may materially and adversely affect an award outstanding under the 2018 Plan without the consent of the affected participant and shareholder approval will be obtained for any amendment to the extent necessary to comply with applicable laws. Further, the plan administrator cannot, without the approval of our shareholders, amend any outstanding share option or SAR to reduce its price per share other than in the context of corporate transactions or equity restructurings, as described above. The 2018 Plan will remain in effect until the tenth anniversary of the earlier of the date that our board of directors or our shareholders approve the 2018 Plan, unless earlier terminated by our board of directors. No awards may be granted under the 2018 Plan after its termination.

Foreign Participants, Claw-Back Provisions, Transferability and Participant Payments

The plan administrator may modify awards granted to participants who are foreign nationals or employed outside the United States or establish subplans or procedures to address differences in laws, rules, regulations or customs of such foreign jurisdictions. All awards will be subject to any company claw-back policy as set forth in such claw-back policy, the 2018 Plan or the applicable award agreement. Except as the plan administrator may determine or provide in an award agreement, awards under the 2018 Plan are generally non-transferrable, except by will or the laws of descent and distribution, or, subject to the plan administrator's consent, pursuant to a domestic relations order, and are generally exercisable only by the participant. With regard to tax withholding obligations arising in connection with awards under the 2018 Plan and exercise price obligations

arising in connection with the exercise of share options under the 2018 Plan, the plan administrator may, in its discretion, accept cash, wire transfer or check, our Class A common shares that meet specified conditions, a promissory note, a "market sell order," such other consideration as the plan administrator deems suitable or any combination of the foregoing.

2018 Employee Share Purchase Plan

Our board of directors adopted and our shareholders have approved, effective the day prior to the first public trading date of our Class A common shares, the 2018 Employee Share Purchase Plan, or the 2018 ESPP. The material terms of the 2018 ESPP are summarized below.

Shares Available for Awards; Administration

A total of 670,000 Class A common shares will initially be reserved for issuance under the 2018 ESPP. In addition, the number of Class A common shares available for issuance under the 2018 ESPP will be annually increased on January 1 of each calendar year beginning in 2019 and ending in and including 2028, by an amount equal to the lesser of (A) 1% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (B) such smaller number of Class A common shares as is determined by our board of directors, provided that no more than 6,420,000 Class A common shares may be issued under the 2018 ESPP. Our board of directors or a committee of our board of directors will administer and will have authority to interpret the terms of a 2018 ESPP and determine eligibility of participants. We expect that the compensation committee will be the initial administrator of the 2018 ESPP.

Eligibility

Our employees are eligible to participate in the 2018 ESPP if they are customarily employed by us or a participating subsidiary for more than twenty hours per week and more than five months in any calendar year. However, an employee may not be granted rights to purchase shares under our 2018 ESPP if the employee, immediately after the grant, would own (directly or through attribution) shares possessing 5% or more of the total combined voting power or value of all classes of our shares.

Grant of Rights

The 2018 ESPP is intended to qualify under Section 423 of the Code and shares will be offered under the 2018 ESPP during offering periods. The length of the offering periods under the 2018 ESPP will be determined by the plan administrator and may be up to twenty-seven months long. Employee payroll deductions will be used to purchase shares on each purchase date during an offering period. The purchase dates for each offering period will be the final trading day in the offering period. Offering periods under the 2018 ESPP will commence when determined by the plan administrator. The plan administrator may, in its discretion, modify the terms of future offering periods.

The 2018 ESPP permits participants to purchase our Class A common shares through payroll deductions of up to a specified percentage of their eligible compensation. The plan administrator will establish a maximum number of shares that may be purchased by a participant during any offering period, which, in the absence of a contrary designation, will be 25,000 shares. In addition, no employee will be permitted to accrue the right to purchase shares under the 2018 ESPP at a rate in excess of $25,000 worth of shares during any calendar year during which such a purchase right is outstanding (based on the fair market value per share of our Class A common shares as of the first day of the offering period).

On the first trading day of each offering period, each participant will automatically be granted an option to purchase our Class A common shares. The option will expire at the end of the applicable offering period, and will be exercised at that time to the extent of the payroll deductions accumulated during the offering period. The purchase price of the shares, in the absence of a contrary designation, will be 85% of the lower of the fair market value of our Class A common shares on the first trading day of the offering period or on the purchase date. Participants may voluntarily end their participation in the 2018 ESPP at any time not later than a specified period prior to the end of the applicable offering period, and will be paid their accrued payroll deductions that have not yet been used to purchase shares of our Class A common shares. Participation ends automatically upon a participant's termination of employment.

A participant may not transfer rights granted under the 2018 ESPP other than by will or the laws of descent and distribution.

Certain Transactions

In the event of certain non-reciprocal transactions or events affecting our Class A common shares, the plan administrator will make equitable adjustments to the 2018 ESPP and outstanding rights. In the event of certain unusual or non-recurring events or transactions, including a change in control, the plan administrator may provide for (1) either the replacement of outstanding rights with other rights or property or termination of outstanding rights in exchange for cash, (2) the assumption or substitution of outstanding rights by the successor or survivor corporation or parent or subsidiary thereof, if any, (3) the adjustment in the number and type of shares subject to outstanding rights, (4) the use of participants' accumulated payroll deductions to purchase shares on a new purchase date prior to the next scheduled purchase date and termination of any rights under ongoing offering periods or (5) the termination of all outstanding rights.

Plan Amendment

The plan administrator may amend, suspend or terminate the 2018 ESPP at any time. However, shareholder approval will be obtained for any amendment that increases the aggregate number or changes the type of shares that may be sold pursuant to rights under the 2018 ESPP, changes the corporations or classes of corporations whose employees are eligible to participate in the 2018 ESPP or changes the 2018 ESPP in any manner that would cause the 2018 ESPP to no longer be an employee share purchase plan within the meaning of Section 423(b) of the Code.

CERTAIN RELATIONSHIPS AND RELATED PERSON TRANSACTIONS

The following includes a summary of transactions since our inception in July 2015 to which we have been a party in which the amount involved exceeded or will exceed $120,000, and in which any of our directors, executive officers or, to our knowledge, beneficial owners of more than 5% of any class of our voting shares or any member of the immediate family of any of the foregoing persons had or will have a direct or indirect material interest, other than equity and other compensation, termination, change in control and other arrangements, which are described under "Executive and Director Compensation." We also describe below certain other transactions with our directors, executive officers and shareholders.

Participation in the Offering

Certain of our existing shareholders, including shareholders affiliated with one of our directors, have indicated an interest in purchasing an aggregate of approximately $50.0 million of Class A common shares in this offering at the initial public offering price per share. However, because indications of interest are not binding agreements or commitments to purchase, the underwriters may determine to sell more, less or no shares in this offering to any of these existing shareholders, or any of these existing shareholders may determine to purchase more, less or no shares in this offering. The underwriters will receive the same underwriting discount on any shares purchased by these existing shareholders as they will on any other shares sold to the public in this offering.

Preferred Share Financings

Series A Preferred Share Financing

In October 2015, we issued and sold an aggregate of 8,028,809 Series A Preferred Shares, and in September 2016 we sold an additional 9,099,311 Series A Preferred Shares to new investors and certain of our directors and executive officers at a price of $4.6707 per share, resulting in aggregate gross proceeds of $80.0 million.

Series B Preferred Share Financing

In March 2017, we issued and sold an aggregate of 5,757,372 Series B Preferred Shares to new investors, existing shareholders, a director and an executive officer at a price of $6.9475 per share, resulting in aggregate gross proceeds of $40.0 million.

Series C Preferred Share Financing

In February 2018, we issued and sold an aggregate of 12,784,601 Series C Preferred Shares to new investors, existing shareholders and certain executive officers at a price of $15.6438 per share, resulting in aggregate gross proceeds of $200.0 million.

The following table sets forth the aggregate number of preferred shares acquired by the listed holders of more than 5% of any class of our voting shares or their affiliated entities and certain of

our executive officers and directors. Each preferred share identified in the following table will convert into one common share upon the closing of this offering.

Preferred Share Participant	Series A	Series B	Series C
5% or Greater Shareholders[1]			
Entities Managed by Baker Bros. Advisors LLP	16,057,618	3,598,392	4,155,000
Arrowpoint Funds	—	575,691	447,458
Cormorant Funds	—	—	830,999
Deerfield Special Situations Fund, L.P	—	—	383,538
Entities Affiliated with Dr. Robert Desnick	585,001	393,282	873,301
Fidelity Funds	—	—	639,230
Sofinnova Venture Partners X L.P.	—	—	894,943
Venrock Funds	—	—	639,230
Officers and Directors[2]			
Sanj K. Patel	428,203	71,967	63,922
Stephen Mahoney	107,050	—	6,392
Thomas Beetham	48,172	—	6,392
Chris Heberlig	74,935	—	6,392
Carsten Boess	107,050	—	6,392
Rasmus Holm-Jorgensen	48,172	—	6,392
Felix J. Baker[3]	16,057,618	3,598,392	4,155,000
Thomas R. Malley[4]	—	71,967	—

(1) Additional details regarding these shareholders and their equity holdings are provided under the caption "Principal Shareholders."

(2) Additional details regarding these officers' and directors' equity holdings are provided under the caption "Principal Shareholders."

(3) Dr. Baker is the beneficial owner of the preferred shares acquired by entities managed by Baker Bros. Advisors LLP. See "Principal Shareholders" for additional details.

(4) Mr. Malley is the beneficial owner of the preferred shares acquired by Mossrock Capital, LLC. See "Principal Shareholders" for additional details.

Our directors, Felix J. Baker, Ph.D. and Stephen R. Biggar, M.D., Ph.D., are associated with 667, L.P. and Baker Brothers Life Sciences, L.P., which beneficially own more than 5% of our Class B common shares.

Investors' Rights Agreement

In connection with our Series C preferred share financing, we entered into an amended and restated investors' rights agreement, or the investors' rights agreement, with holders of our preferred shares, including certain executive officers, holders of 5% of any class of our voting shares and entities affiliated with certain of our directors. The investors' rights agreement, among other things, grants these shareholders specified registration rights with respect to our Class A common shares, including common shares issued or issuable upon conversion of the preferred shares held by them. For more information regarding the registration rights provided in these agreements, please refer to the section entitled "Description of Share Capital — Registration Rights."

Voting Agreement

In connection with our Series C preferred share financing, we entered into a second amended and restated voting agreement in February 2018, or the voting agreement, with holders of our preferred shares, including certain executive officers, holders of 5% of our any class of our voting shares and entities affiliated with certain of our directors. Pursuant to the voting agreement, the preferred shareholders agreed to elect the following directors to serve as members on our board of

directors: Sanj K. Patel, Felix J. Baker, Ph.D., Stephen R. Biggar, M.D., Ph.D., Barry D. Quart, Pharm.D., Thomas R. Malley, Tracey L. McCain and Kimberly J. Popovits. As of the date of this prospectus, each of these directors continues to serve on our board of directors. Pursuant to the voting agreement, Drs. Baker and Biggar were initially selected to serve on our board of directors as the directors designated by the holders of our preferred shares. Mr. Patel was initially selected to serve on our board of directors in his capacity as our Chief Executive Officer. Dr. Quart, Mr. Malley and Mses. McCain and Popovits were initially selected to serve on our board of directors as independent directors. The voting agreement will terminate upon the closing of this offering, and members previously elected to our board of directors pursuant to this agreement will continue to serve as directors until they have completed their term.

Indemnification Agreements

We have entered into indemnification agreements with all of our directors and executive officers. These agreements, among other things, require us or will require us to indemnify each director (and in certain cases their related investment funds) and executive officer to the fullest extent permitted by Bermuda law, including indemnification of expenses such as attorneys' fees, judgments, fines and settlement amounts incurred by the director or executive officer in any action or proceeding, including any action or proceeding by or in right of the Company, arising out of such person's services as a director or executive officer.

Employment Agreements

In connection with this offering, we intend to amend the existing employment agreements with our named executive officers. The material terms of those arrangements are described in "Executive and Director Compensation — Executive Compensation Arrangements." In addition, we intend to amend existing employment agreements with certain of our officers as described below.

Chris Heberlig

Chris Heberlig serves as our Executive Vice President and Chief Financial Officer. Mr. Heberlig's employment agreement will provide for an annual base salary of $350,000, which may be changed from time to time in the discretion of our board of directors. Mr. Heberlig will be eligible to earn an annual discretionary cash bonus with a target of 35% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Thomas Beetham

Thomas Beetham serves as our Executive Vice President and Chief Legal Officer. Mr. Beetham's employment agreement will provide for an annual base salary of $375,000, which may be changed from time to time in the discretion of our board of directors. Mr. Beetham will be eligible to earn an annual discretionary cash bonus with a target of 35% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Carsten Boess

Carsten Boess serves as our Executive Vice President, Corporate Affairs. Mr. Boess' employment agreement will provide for an annual base salary of $400,000, which may be changed from time to time in the discretion of our board of directors. Mr. Boess will be eligible to earn an annual discretionary cash bonus with a target of 35% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Rasmus Holm-Jorgensen

Rasmus Holm-Jorgensen serves as our Senior Vice President, Chief Strategy and Portfolio Officer. Mr. Holm-Jorgensen's employment agreement will provide for an annual base salary of $337,740, which may be changed from time to time in the discretion of our board of directors. Mr. Holm-Jorgensen will be eligible to earn an annual discretionary cash bonus with a target of 30% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Share Option Grants to Officers and Directors

We have granted share options for our Class A common shares to our named executive officers and certain of our directors as more fully described in the section entitled "Executive and Director Compensation." In December 2015, we granted 125,941 share options each to Messrs. Heberlig, Beetham, and Boess and 75,565 share options to Mr. Holm-Jorgensen, each at an exercise price of $1.59 per share. In June 2017, we granted 62,475 share options to Mr. Heberlig, 66,135 share options to Mr. Beetham, 36,598 share options to Mr. Boess, and 40,689 share options to Mr. Holm-Jorgensen, each at an exercise price of $3.80. In March 2018, we granted 109,795 share options each to Messrs. Heberlig and Beetham, 9,149 share options to Mr. Boess, and 36,598 share options to Mr. Holm-Jorgensen, each at an exercise price of $10.36.

Policies and Procedures for Related Person Transactions

Our board of directors adopted a written related person transaction policy, to be effective upon the effectiveness of the registration statement of which this prospectus forms a part, setting forth the policies and procedures for the review and approval or ratification of related person transactions. This policy will cover, with certain exceptions set forth in Item 404 of Regulation S-K under the Securities Act, any transaction, arrangement or relationship, or any series of similar transactions, arrangements or relationships, in which we were or are to be a participant, where the amount involved exceeds $120,000 in any fiscal year and a related person had, has or will have a direct or indirect material interest, including without limitation, purchases of goods or services by or from the related person or entities in which the related person has a material interest, indebtedness, guarantees of indebtedness and employment by us of a related person. In reviewing and approving any such transactions, our audit committee considers all relevant facts and circumstances, including, but not limited to, whether the transaction is on terms comparable to those that could be obtained in an arm's length transaction and the extent of the related person's interest in the transaction. All of the transactions described in this section occurred prior to the adoption of this policy.

PRINCIPAL SHAREHOLDERS

The following table sets forth information with respect to the beneficial ownership of our Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares, as of March 31, 2018, and as adjusted to reflect the sale of Class A common shares in this offering, by:

- each person or group of affiliated persons known by us to beneficially own more than 5% of our Class A common shares or Class B common shares;
- each of our named executive officers and directors; and
- all of our executive officers and directors as a group.

The number of shares beneficially owned by each shareholder is determined under rules issued by the SEC. Under these rules, beneficial ownership includes any shares over which the individual or entity has sole or shared voting power or investment power. Applicable percentage ownership is based on 6,270,569 Class A common shares outstanding, 12,995,954 Class A1 common shares outstanding, 4,638,855 Class B common shares outstanding, and 16,057,618 Class B1 common shares outstanding, each as of March 31, 2018, assuming the conversion of all outstanding preferred shares into 5,546,019 Class A common shares, 12,995,954 Class A1 common shares, 1,070,502 Class B common shares, and 16,057,618 Class B1 common shares in connection with this offering. In computing the number of shares beneficially owned by an individual or entity and the percentage ownership of that person, common shares subject to options or other rights held by such person that are currently exercisable or will become exercisable within 60 days of March 31, 2018 are considered outstanding, although these shares are not considered outstanding for purposes of computing the percentage ownership of any other person.

Unless noted otherwise, the address of all listed shareholders is c/o Kiniksa Pharmaceuticals Corp., 100 Hayden Avenue, Lexington, Massachusetts 02421. Each of the shareholders listed has sole voting and investment power with respect to the shares beneficially owned by the shareholder unless noted otherwise, subject to community property laws where applicable.

Certain of our existing shareholders, including shareholders affiliated with one of our directors, have indicated an interest in purchasing an aggregate of approximately $50.0 million of Class A common shares in this offering at the initial public offering price per share. However, because indications of interest are not binding agreements or commitments to purchase, the underwriters may determine to sell more, less or no shares in this offering to any of these existing shareholders, or any of these existing shareholders may determine to purchase more, less or no shares in this offering. The underwriters will receive the same underwriting discount on any shares purchased by these existing shareholders as they will on any other shares sold to the public in this offering. The following table does not reflect any such purchases by these existing shareholders or their affiliated entities.

Our Class A1 common shares are convertible into Class A common shares at any time at the option of the holder, with prior notice to us, on a one-for-one basis, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of the issued and outstanding Class A common shares. Accordingly, each holder of Class A1 common shares is deemed to be the beneficial owner of the number of Class A common shares that would result in such holder owning up to 4.99% of the issued and outstanding Class A common shares, in addition to any other Class A common shares beneficially owned by such holder.

Our Class B common shares are convertible into Class A common shares or Class B1 common shares at any time at the option of the holder, with prior notice to us, on a one-for-one

basis. Accordingly, each holder of Class B common shares is deemed to be the beneficial owner of, in each case, an equal number of Class A common shares and Class B1 common shares, in addition to any other Class A common shares or Class B1 common shares beneficially owned by such holder.

Our Class B1 common shares are convertible into Class A common shares or Class B common shares at any time at the option of the holder, with prior notice to us, on a one-for-one basis, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of the issued and outstanding Class A common shares. Accordingly, each holder of Class B1 common shares is deemed to be the beneficial owner of the number of Class A common shares and Class B common shares, in each case, that would result in such holder owning up to 4.99% of the issued and outstanding Class A common shares, in addition to any other Class A common shares or Class B common shares beneficially owned by such holder.

	Beneficial Ownership Before the Offering									Beneficial Ownership After the Offering								
	Class A common shares		Class A1 common shares		Class B common shares		Class B1 common shares		% of Total Voting Power Before the Offering	Class A common shares		Class A1 common shares		Class B common shares		Class B1 common shares		% of Total Voting Power After the Offering
	Shares	%	Shares	%	Shares	%	Shares	%		Shares	%	Shares	%	Shares	%	Shares	%	
Name of Beneficial Owner:																		
5% or Greater Shareholders:																		
Arrowpoint Funds[1]	1,020,408	16.27%	—	—	—	—	—	—	1.94%	1,020,408	7.69%	—	—	—	—	—	—	1.71%
Sofinnova Venture Partners X, L.P.[2]	894,923	14.27%	—	—	—	—	—	—	1.70%	894,923	6.74%	—	—	—	—	—	—	1.50%
Cormorant Funds[3]	830,999	13.25%	—	—	—	—	—	—	1.58%	830,999	6.26%	—	—	—	—	—	—	1.39%
Entities Affiliated with Dr. Robert Desnick[4]	679,631	10.84%	—	—	214,101	4.62%	214,101	1.03%	4.95%	679,631	5.04%	—	—	214,101	4.62%	214,101	1.03%	4.37%
Fidelity Funds[5]	639,230	10.19%	—	—	—	—	—	—	1.21%	639,230	4.82%	—	—	—	—	—	—	1.07%
Venrock Funds[6]	639,230	10.19%	—	—	—	—	—	—	1.21%	639,230	4.82%	—	—	—	—	—	—	1.07%
Deerfield Special Situations Fund, L.P.[7]	383,538	6.12%	—	—	—	—	—	—	*	383,538	2.89%	—	—	—	—	—	—	*
Entities managed by Baker Bros. Advisors LP[8]	328,988	4.99%	7,753,392	59.66%	328,988	6.62%	16,057,618	77.59%	—	696,246	4.99%	7,753,392	59.66%	696,246	13.05%	16,057,618	77.59%	—
Officers and Directors:																		
Sanj K. Patel[9]	2,227,979	28.88%	—	—	1,526,160	32.90%	1,526,160	7.37%	29.66%	2,227,979	14.71%	—	—	1,526,160	32.90%	1,526,160	7.37%	26.18%
Thomas Beetham[10]	580,330	8.63%	—	—	414,157	8.93%	414,157	2.00%	8.02%	580,330	4.21%	—	—	414,157	8.93%	414,157	2.00%	7.08%
Carsten Boess[11]	639,208	9.51%	—	—	473,035	10.20%	473,035	2.29%	9.13%	639,208	4.62%	—	—	473,035	10.20%	473,035	2.29%	8.06%
Chris Heberlig[12]	607,093	9.03%	—	—	440,920	9.50%	440,920	2.13%	8.52%	607,093	4.40%	—	—	440,920	9.50%	440,920	2.13%	7.52%
Rasmus Holm-Jorgensen[13]	457,868	6.92%	—	—	340,960	7.35%	340,960	1.65%	6.60%	457,868	3.35%	—	—	340,960	7.35%	340,960	1.65%	5.82%
Stephen Mahoney[14]	858,798	12.44%	—	—	656,027	14.14%	656,027	3.17%	12.68%	858,798	6.13%	—	—	656,027	14.14%	656,027	3.17%	11.19%
John F. Paolini, M.D., Ph.D.[15]	99,563	1.56%	—	—	—	—	—	—	*	99,563	*	—	—	—	—	—	—	*
Felix J. Baker, Ph.D.[8]	328,988	4.99%	7,753,392	59.66%	328,988	6.62%	16,057,618	77.59%	—	696,246	4.99%	7,753,392	59.66%	696,246	13.05%	16,057,618	77.59%	—
Stephen R. Biggar, M.D., Ph.D.	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	
Thomas R. Malley[16]	86,604	1.38%	—	—	—	—	—	—	*	86,604	*	—	—	—	—	—	—	*
Tracey L. McCain	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—
Kimberly J. Popovits	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—
Barry D. Quart, Pham. D.[17]	14,181	*	—	—	—	—	—	—	*	14,181	*	—	—	—	—	—	—	*
All executive officers and directors as a group (13 persons)[18]	**4,803,537**	**47.83%**	**7,753,392**	**59.66%**	**3,037,264**	**65.47%**	**16,057,618**	**77.59%**	**59.01%**	**5,170,795**	**30.34%**	**7,753,392**	**59.66%**	**3,037,264**	**65.47%**	**16,057,618**	**77.59%**	**52.09%**

* Represents beneficial ownership less than 1%

(1) Consists of (a) 173,010 Class A common shares held directly by ArrowMark Fundamental Opportunity Fund, L.P. ("Opportunity Fund"), (b) 513,854 Class A common shares held directly by Meridian Small Cap Growth Fund ("Meridian"), (c) 52,034 Class A common shares held directly by Lookfar Investments LLC ("Lookfar"), (d) 93,490 common shares held directly by Iron Horse Investments LLC ("Iron Horse"), (e) 185,279 common shares held directly by THB Iron Rose LLC ("Rose"), and (f) 2,741 Class A common shares held directly by THB Iron Rose LLC, Life Science Portfolio ("Life Science") (collectively, the "ArrowMark Funds"). ArrowMark Partners GP, LLC ("ArrowMark GP") is the general partner of Opportunity Fund and David Corkins is the managing member of ArrowMark GP. ArrowMark Colorado Holdings LLC ("ArrowMark Colorado") is investment advisor to Meridian, Lookfar, Iron Horse, Rose and Life Science (collectively, the "ArrowMark Colorado Funds"). Mr. Corkins is a managing member of ArrowMark Colorado and Mr. Yao is a portfolio manager of ArrowMark Colorado. Mr. Corkins may be considered the beneficial owner of the shares held by the ArrowMark Funds and Mr. Yao may be considered the beneficial owner of the shares held by ArrowMark Colorado. The principal business address of the ArrowMark Funds is 100 Fillmore Street, Suite 325, Denver, Colorado 80206.

(2) Sofinnova Management IX, L.L.C. ("Sofinnova Management") is the general partner of Sofinnova Venture Partners X, L.P. ("Sofinnova X") and Anand Mehra, Michael Powell and James Healy are the managing members of Sofinnova Management. Sofinnova Management, Anand Mehra (a member of our board), Michael Powell and James Healy may be deemed to beneficially own the shares owned by Sofinnova X. Such entities and individuals disclaim beneficial ownership over all shares except to the extent of any pecuniary interest therein. The address for Sofinnova X and Sofinnova Management is 3000 Sand Hill Road, Building 4, Suite 250, Menlo Park, California 94025.

(3) Consists of (a) 640,617 Class A common shares held directly by Cormorant Private Healthcare Fund I, L.P. ("Cormorant I"), (b) 161,962 Class A common shares held directly by Cormorant Global Healthcare Master Fund, LP ("Cormorant Master Fund"); and (c) 28,420 Class A common shares held directly by CRMA SPV, L.P. ("CRMA" and with Cormorant I and Cormorant Master Fund, the "Cormorant Funds"). Cormorant Global Healthcare GP, LLC ("Global GP") is the general partner of Cormorant Master Fund, Cormorant Private Healthcare GP, LLC ("Private GP") is the general partner of Cormorant I. Bihua Chen is the sole managing member of both Global GP and Private GP. Cormorant Asset Management LLP ("Cormorant Management") serves as the investment manager to CRMA, and Ms. Chen is the sole managing member of Cormorant Asset Management GP, LLC. Ms. Chen may be deemed to beneficially own the shares held by the Cormorant Funds. The address of the Cormorant Funds, Global GP, Private GP, Cormorant Management, and Ms. Chen is 200 Clarendon Street, 52nd Floor, Boston, MA 02116.

(4) Consists of (a) 384,666 Class A common shares held by Edward Schuchman, Ph.D., as the trustee for the Desnick / Herzig 2012 GST Trust UAD 10/32/12 ("Trust") and New Direction IRA, Inc. FBO Robert Desnick Roth IRA ("New Direction"), (b) 214,101 Class B common shares held directly by New Direction and (c) 3,659 Class A common shares that Dr. Desnick has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options. Dr. Desnick directs the voting and investment of the shares held by Trust and New Direction and may be deemed to beneficially own the shares owned by Trust and New Direction.

(5) Consists of (a) 304,279 Class A common shares held by Fidelity Mt. Vernon Street Trust: Fidelity Growth Company Fund, whose address is 525 William Penn Place Run Rm 040, Pittsburgh, PA 15259, ("Fidelity Growth Fund"), (b) 244,7213 Class A common shares held by Fidelity Growth Company Commingled Pool, whose address is c/o Brown Brothers Harriman & Co., 140 Broadway, New York, NY 10005, ("Fidelity Commingled Fund"), and (c) 90,238 Class A common shares held by Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund, whose address is PO Box 5756, Boston, MA 02206 (together with Fidelity Growth Fund and Fidelity Commingled Fund, the "Fidelity Growth Funds"). These accounts are managed by direct or indirect subsidiaries of FMR LLC. Abigail P. Johnson is a Director, the Vice Chairman, the Chief Executive Officer and the President of FMR LLC. Members of the Johnson family, including Abigail P. Johnson, are the predominant owners, directly or through trusts, of Series B voting common shares of FMR LLC, representing 49% of the voting power of FMR LLC. The Johnson family group and all other Series B shareholders have entered into a shareholders' voting agreement under which all Series B voting common shares will be voted in accordance with the majority vote of Series B voting common shares. Accordingly, through their ownership of voting common shares and the execution of the shareholders' voting agreement, members of the Johnson family may be deemed, under the Investment Company Act of 1940, to form a controlling group with respect to FMR LLC. Neither FMR LLC nor Abigail P. Johnson has the sole power to vote or direct the voting of the shares owned directly by the various investment companies registered under the Investment Company Act ("Fidelity Funds") advised by Fidelity Management & Research Company ("FMR Co"), a wholly owned subsidiary of FMR LLC, which power resides with the Fidelity Funds' Boards of Trustees. Fidelity Management & Research Company carries out the voting of the shares under written guidelines established by the Fidelity Funds' Boards of Trustees.

(6) Consists of (a) 454,812 Class A common shares held directly by Venrock Healthcare Capital Partners II, L.P. ("VHCP II") and (b) 184,418 Class A common shares held directly by VHCP Co-Investment Holdings II, LLC ("Co-Invest II"). VHCP Management II, LLC ("VHCP Management II") is the general partner of VHCP II and the manager of Co-Invest II and may be deemed to beneficially own these shares. Drs. Bong Koh and Nimish Shah are the managing members of VHCP Management II and may be deemed to beneficially own the shares. Drs. Koh and Shah and VHCP Management II expressly disclaim beneficial ownership over the shares except to the extent of their indirect pecuniary interests therein. The address for VHCP II, Co-Invest and Drs. Koh and Shah is 530 Fifth Avenue, 22nd Floor, New York, NY 10036.

(7) Deerfield Mgmt, L.P. is the general partner of Deerfield Special Situations Fund, L.P. (the "Fund"). Deerfield Management Company, L.P. is the investment manager of the Fund. James E. Flynn is the sole member of the general partner of each of Deerfield Mgmt, L.P. and Deerfield Management Company, L.P. Deerfield Mgmt, L.P., Deerfield Management and Mr. Flynn may be deemed to be the beneficial owners of the shares owned by Fund. Deerfield Mgmt, L.P., Deerfield Management Company, L.P. and Mr. Flynn disclaim beneficial ownership of such shares except to the extent of its or his pecuniary interest therein. The address for the Fund is 780 Third Avenue, 37th Floor, New York, NY 10017.

(8) Consists of (a) 7,018,874 Class A1 common shares held directly by Baker Brothers Life Sciences, L.P. ("BBLS"), (b) 734,581 Class A1 common shares held by 667, L.P. ("667" and with BBLS, the "Baker Funds"), (c) 14,658,102 Class B1 common shares held directly by BBLS, and (d) 1,399,516 Class B1 common shares held directly by 667. Baker Bros. Advisors LP ("Advisors") is the management company and investment advisor to BBLS and 667. Felix J. Baker is a managing member of Advisors and may be deemed to beneficially own the shares owned by the Baker Funds. Dr. Baker disclaims beneficial ownership of such shares except to the extent of any pecuniary interest therein. The address for the Baker Funds is 860 Washington Street, 3rd Floor, New York, NY 10014 .

(9) Consists of (a) 245,685 Class A common shares held directly by Mr. Patel and 109,795 Class A common shares held by Mr. Patel as trustee for the Manisha S. Patel 2016 Irrevocable Trust, (b) 1,526,160 Class B common shares, and (c) 346,339 Class A common shares that Mr. Patel has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(10) Consists of (a) 79,789 Class A common shares, (b) 414,157 Class B common shares and (c) 86,584 Class A common shares that Mr. Beetham has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(11) Consists of (a) 79,589 Class A common shares, (b) 473,035 Class B common shares, and (c) 86,584 Class A common shares that Mr. Boess has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(12) Consists of (a) 42,991 Class A common shares held directly by Mr. Heberlig and 36,598 Class A common shares held by Sandra C. Heberlig, Mr. Heberlig's spouse, as trustee for the Christopher J. Heberlig 2017 Irrevocable Trust, (b) 440,920 Class B common shares, and (c) 86,584 Class A common shares that Mr. Heberlig has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(13) Consists of (a) 64,949 Class A common shares, (b) 340,960 Class B common shares, and (c) 51,959 Class A common shares that Mr. Holm-Jorgensen has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(14) Consists of (a) 79,589 Class A common shares held directly by Mr. Mahoney and 36,598 Class A common shares held by Krisha S. Mahoney, Mr. Mahoney's spouse, as trustee for the Stephen F. Mahoney 2016 Irrevocable Trust, (b) 656,027 Class B common shares, and (c) 86,584 Class A common shares that Mr. Mahoney has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(15) Includes 99,563 Class A common shares that Dr. Paolini has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(16) Consists of (a) 71,967 common shares held by Mossrock Capital, LLC ("Mossrock") and (b) 14,637 Class A common shares that Mr. Malley has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options. Mr. Malley is the president of Mossrock and may be deemed to beneficially own the shares owned by Mossrock. The address of Mossrock is 19 Martin Lane, Englewood, CO 80113.

(17) Includes 14,181 Class A common shares that Dr. Quart has the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

(18) Consists of (a) 702,812 Class A common shares, (b) 7,753,392 Class A1 common shares, (c) 3,037,264 Class B common shares, (d)16,057,618 Class B1 common shares, and (e) 734,473 Class A common shares that all officers and directors as a group have the right to acquire within 60 days following March 31, 2018 pursuant to the exercise of share options.

DESCRIPTION OF SHARE CAPITAL

The following description of our share capital and provisions of our memorandum of association and amended and restated bye-laws are summaries. You should also refer to our memorandum of association and amended and restated bye-laws, which are filed as exhibits to the registration statement of which this prospectus is part.

General

We are an exempted company incorporated under the laws of Bermuda. We are registered with the Registrar of Companies in Bermuda under registration number 50484. We were incorporated on July 21, 2015. Our registered office is located in Bermuda at Clarendon House, 2 Church Street, Hamilton HM11, Bermuda.

The objects of our business are unrestricted, and we have the capacity of a natural person. We can therefore undertake activities without restriction on our capacity.

Our board of directors and shareholders have approved certain amendments to our bye-laws that will become effective immediately following the closing of this offering. The following description assumes that such amendments have become effective.

Since our incorporation, other than a subdivision of our designated and issued share capital, there have been no material changes to our share capital, mergers, amalgamations or consolidations of us or any of our subsidiaries, no material changes in the mode of conducting our business, no material changes in the types of products produced or services rendered. There have been no bankruptcy, receivership or similar proceedings with respect to us or our subsidiaries.

There have been no public takeover offers by third parties for our shares nor any public takeover offers by us for the shares of another company that have occurred during the last or current financial years.

Share Capital

Following this offering, our authorized share capital will consist of 200,000,000 shares, par value $0.000273235 per share, and there will be 13,270,569 Class A common shares, 4,638,855 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares issued and outstanding.

Pursuant to our amended and restated bye-laws, subject to the requirements of Nasdaq and subject to any resolution of the shareholders to the contrary, our board of directors is authorized to issue any of our designated but unissued shares. There are no limitations on the right of non-Bermudians or non-residents of Bermuda to hold or vote our shares.

Common Shares

Following this offering, we will have four classes of shares: Class A, Class A1, Class B and Class B1. Class A and Class B common shares are voting common shares, or together the voting common shares, and Class A1 and Class B1 are non-voting common shares. Except as described in this prospectus with respect to voting rights conversion and transferability each common share will have the same rights and powers of, rank equally to, share ratably with and will be identical in all respects and as to all matters with each other common share. In the event of our liquidation, dissolution or winding up, the holders of our common shares are entitled to share equally and ratably in our assets, if any, remaining after the payment of all of our debts and liabilities, subject to any liquidation preference on any issued and outstanding preferred shares. None of our common shares have pre-emptive, redemption or sinking fund rights.

Class A Common Shares

The shares being offered in this offering are our Class A common shares. As of March 31, 2018, there were 724,550 Class A common shares issued and outstanding. All Class A common shares are fully paid and non-assessable.

Class B Common Shares

As of March 31, 2018, there were 3,568,353 Class B common shares outstanding. Each holder of Class B common shares may convert any portion of its Class B common shares into Class A common shares or Class B1 common shares at any time. In addition, each Class B common share automatically converts into one Class A common share upon transfer, except for transfers to or between affiliated holders. Our Class B common shares also have greater voting power than our Class A common shares, as described in "— Voting Rights."

Class A1 Common Shares

As of March 31, 2018, there were no Class A1 common shares issued and outstanding. No Class A1 common shares may be issued until the effectiveness of the registration statement of which this prospectus forms a part. Following this offering, each holder of Class A1 common shares may elect to convert any portion of its Class A1 common shares into voting Class A common shares at any time, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own or would beneficially own more than 4.99% of the issued and outstanding Class A common shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act. A holder of Class A1 common shares may increase, decrease or waive this limitation on ownership by providing us with 61-days' notice. In addition, each Class A1 common share automatically converts into one Class A common share upon transfer, except for transfers to or between affiliated holders.

Class B1 Common Shares

As of March 31, 2018, there were no Class B1 common shares issued and outstanding. No Class B1 common shares may be issued until the effectiveness of the registration statement of which this prospectus forms a part. Following this offering, each holder of Class B1 common shares may elect to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own or would beneficially own more than 4.99% of the issued and outstanding Class A common shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act. A holder of Class B1 common shares may increase, decrease or waive this limitation on ownership by providing us with 61-days' notice. In addition, each Class B1 common share automatically converts into one Class A common share upon transfer, except for transfers to or between affiliated holders.

Preferred Shares

Under Bermuda law and our amended and restated bye-laws, our board of directors is authorized to issue preferred shares in one or more series without shareholder approval. Our board of directors has the discretion under the bye-laws to determine the rights, preferences, privileges and restrictions, including voting rights, dividend rights, conversion rights, redemption privileges and liquidation preferences, of each series of preferred shares, without any further shareholder approval. The rights with respect to a series of preferred shares may be greater than the rights attached to our Class A common shares. It is not possible to state the actual effect of the issuance of any preferred shares on the rights of holders of our common shares until our board of directors

determines the specific rights attached to those preferred shares. The effect of issuing preferred shares could include, among other things, one or more of the following:

- restricting dividends in respect of our Class A common shares;
- diluting the voting power of our Class A common shares or providing that holders of preferred shares have the right to vote on matters as a class;
- impairing the liquidation rights of our common shares; or
- delaying or preventing a change of control of us.

Upon the consummation of this offering, there will be no preferred shares outstanding, and we have no present plans to issue any preferred shares following the offering.

Voting Rights

Unless a different majority is required by Bermuda law or by our amended and restated bye-laws, resolutions to be approved by holders of voting common shares require approval by a simple majority of votes cast at a meeting at which a quorum is present. Holders of our voting common shares vote together as a single class on all matters presented to the shareholders for their vote or approval, including the election of directors. Any individual who is a shareholder and who is present and entitled to vote at a meeting may vote in person, as may any corporate shareholder that is represented by a duly authorized representative at a meeting of shareholders. Our amended and restated bye-laws also permit attendance at general meetings by proxy, provided the instrument appointing the proxy is in the form specified in the bye-laws or such other form as the board of directors may determine.

Each Class A common share is entitled to one vote per share and each Class B common share is entitled to ten votes per share. Each Class A1 common share and Class B1 common share is non-voting. Immediately following this offering, the holders of Class A common shares will account for 22.2% of our aggregate voting power and the holders of Class B common shares will account for the remaining 77.8% of our aggregate voting power. Our bye-laws will generally provide that holders of our voting common shares are entitled to vote, on a non-cumulative basis, at all annual general and special general meetings of shareholders with respect to matters on which voting common shares are eligible to vote. However, these percentages may change depending on any conversion of Class A1 and Class B1 common shares into voting common shares, to the extent any are issued, and any conversion of Class B common shares into Class A common shares. See " — Common Shares" for more information.

Dividend Rights

Under Bermuda law and our amended and restated bye-laws, we may not declare or pay dividends if there are reasonable grounds for believing that: (i) we would after the payment be unable to pay our liabilities as they become due; or (ii) that the realizable value of our assets would thereby be less than our liabilities. Under our bye-laws, each common share is entitled to dividends if, as and when dividends are declared by our board of directors, subject to any preferred dividend right of the holders of any preferred shares. There are no restrictions on our ability to transfer funds (other than funds denominated in Bermuda dollars) in and out of Bermuda or to pay dividends to U.S. residents who are holders of our common shares.

We intend to retain future earnings, if any, to finance the operation and expansion of our business and do not anticipate paying any cash dividends on our Class A common shares in the foreseeable future. Any future determination related to our dividend policy will be made at the

discretion of our board of directors after considering our financial condition, results of operations, capital requirements, business prospects and other factors the board of directors deems relevant.

We are a holding company and have no direct operations. As a result, we will depend upon distributions from our subsidiaries to pay any dividends.

Variation of Rights

If at any time we have more than one class of shares, the rights attaching to any class, unless otherwise provided for by the terms of issue of the relevant class, may be varied either: (i) with the consent in writing of the holders of 75% of the issued shares of that class; or (ii) with the sanction of a resolution passed by a majority of the votes cast at a general meeting of the relevant class of shareholders at which a quorum consisting of at least two persons holding or representing one-third of the issued shares of the relevant class is present. Our amended and restated bye-laws specify that the creation or issue of shares ranking equally with existing shares or the purchase or redemption by us of our shares will not, unless expressly provided by the terms of issue of existing shares, vary the rights attached to existing shares. In addition, the creation or issue of preferred shares ranking prior to common shares will not be deemed to vary the rights attached to common shares or, subject to the terms of any other series of preferred shares, to vary the rights attached to any other series of preferred shares.

Transfer of Shares

Our board of directors may in its absolute discretion and without assigning any reason refuse to register the transfer of a share that it is not fully paid. The board of directors may also refuse to recognize an instrument of transfer of a share unless it is accompanied by the relevant share certificate and such other evidence of the transferor's right to make the transfer as the board of directors shall reasonably require. Subject to these restrictions, a holder of common shares may transfer the title to all or any of such holder's common shares by completing a form of transfer in the form set out in the bye-laws (or as near thereto as circumstances admit) or in such other common form as the board of directors may accept. The instrument of transfer must be signed by the transferor and transferee, although in the case of a fully paid share the board of directors may accept the instrument signed only by the transferor.

Meetings of Shareholders

Under the Companies Act, a company is required to convene at least one general meeting of shareholders each calendar year, which is referred to as the annual general meeting. However, the members may by resolution waive this requirement, either for a specific year or period of time, or indefinitely. When the requirement has been so waived, any member may, on notice to the company, terminate the waiver, in which case an annual general meeting must be called.

The Companies Act provides that a special general meeting of shareholders may be called by the board of directors of a company and must be called upon the request of shareholders holding not less than 10% of the paid-up capital of the company carrying the right to vote at general meetings. The Companies Act also requires that shareholders be given at least five days' advance notice of a general meeting, but the accidental omission to give notice to any person does not invalidate the proceedings at a meeting. Our bye-laws provide that our President or Chairman or any two directors or any director and secretary may convene an annual general meeting or a special general meeting. Under our bye-laws, at least twenty days' notice of an annual general meeting or a special general meeting must be given to each shareholder entitled to vote at such meeting. This notice requirement is subject to the ability to hold such meetings on shorter notice if such notice is agreed: (i) in the case of an annual general meeting by all of the shareholders

entitled to attend and vote at such meeting; or (ii) in the case of a special general meeting by a majority in number of the shareholders entitled to attend and vote at the meeting holding not less than 95% in nominal value of the shares entitled to vote at such meeting. The quorum required for a general meeting of shareholders is two or more persons present throughout the meeting and representing in person or by proxy a majority of the voting power of the issued and outstanding voting shares.

Access to Books and Records and Dissemination of Information

Members of the general public have a right to inspect the public documents of a company available at the office of the Registrar of Companies in Bermuda. These documents include a company's memorandum of association, including its objects and powers, and certain alterations to the memorandum of association. The shareholders have the additional right to inspect the bye-laws of a company, minutes of general meetings and a company's audited financial statements, which must be presented to the annual general meeting. The register of members of a company is also open to inspection by shareholders and by members of the general public without charge. The register of members is required to be open for inspection for not less than two hours in any business day (subject to the ability of a company to close the register of members for not more than thirty days in a year). A company is required to maintain its share register in Bermuda but may, subject to the provisions of the Companies Act, establish a branch register outside of Bermuda. A company is required to keep at its registered office a register of directors and officers that is open for inspection for not less than two hours in any business day by members of the public without charge. Bermuda law does not, however, provide a general right for shareholders to inspect or obtain copies of any other corporate records.

Election and Removal of Directors

Our amended and restated bye-laws provide that our board of directors shall consist of not less than five members and not more than such number of directors as the board of directors determine. Upon the closing of this offering, our board of directors will consist of seven directors. Our board of directors will be divided into three classes that are, as nearly as possible, of equal size. Each class of directors will be elected for a three-year term of office, but the terms will be staggered so that the term of only one class of directors expires at each annual general meeting. Following the election of all of the directors at the first general meeting of shareholders following this offering, the initial terms of the Class I, Class II and Class III directors will expire in 2022, 2020 and 2021, respectively. At each succeeding annual general meeting, successors to the class of directors whose term expires at the annual general meeting will be elected for a three-year term.

A shareholder holding any percentage of the common shares in issue may propose for election as a director someone who is not an existing director or is not proposed by our board of directors. Where a director is to be elected at an annual general meeting, notice of any such proposal for election must be given not less than 90 days nor more than 120 days before the anniversary of the last annual general meeting prior to the giving of the notice or, in the event the annual general meeting is called for a date that is not less than 30 days before or after such anniversary the notice must be given not later than ten days following the earlier of the date on which notice of the annual general meeting was posted to shareholders or the date on which public disclosure of the date of the annual general meeting was made. Where a director is to be elected at a special general meeting; provided, that our board of directors has determined that shareholders may nominate persons for election at such special general meeting, that notice must be given not later than seven days following the earlier of the date on which notice of the special general meeting was posted to shareholders or the date on which public disclosure of the date of the special general meeting was made.

A director may be removed, only with cause, by the shareholders, provided notice of the shareholders meeting convened to remove the director is given to the director. The notice must contain a statement of the intention to remove the director and a summary of the facts justifying the removal and must be served on the director not less than 14 days before the meeting. The director is entitled to attend the meeting and be heard on the motion for his removal.

Proceedings of Board of Directors

Our amended and restated bye-laws provide that our business is to be managed and conducted by our board of directors. Bermuda law permits individual and corporate directors, and there is no requirement in our amended and restated bye-laws or Bermuda law that directors hold any of our shares. There is also no requirement in our amended and restated bye-laws or under Bermuda law that our directors must retire at a certain age.

The remuneration of our directors is determined by the board of directors and each such director, other than directors who are employees of the Company, shall be paid a fee at a rate determined by the board of directors. The directors may also be paid all travel, hotel and other expenses properly incurred by them in connection with our business or their duties as directors.

A director who has a direct or indirect interest in any contract or arrangement with the Company must disclose such interest as required by the Companies Act. Such an interested director is not entitled to vote on or participate in any discussion in respect of any such contract or arrangement in which he or she is interested.

Indemnification of Directors and Officers

Section 98 of the Companies Act provides generally that a Bermuda company may indemnify its directors, officers and auditors against any liability which by virtue of any rule of law would otherwise be imposed on them in respect of any negligence, default, breach of duty or breach of trust, except in cases where such liability arises from fraud or dishonesty of which such director, officer or auditor may be guilty in relation to the Company. Section 98 further provides that a Bermuda company may indemnify its judgment is awarded in their favor or in which they are acquitted or granted relief by the Supreme Court of Bermuda pursuant to section 281 of the Companies Act.

We have adopted provisions in our amended and restated bye-laws that provide that we shall indemnify our officers and directors in respect of their actions and omissions, except in respect of their fraud or dishonesty. Our amended and restated bye-laws provide that the shareholders waive all claims or rights of action that they might have, individually or in right of the Company, against any of the Company's directors or officers for any act or failure to act in the performance of such director's or officer's duties, except in respect of any fraud or dishonesty of such director or officer. Section 98A of the Companies Act permits us to purchase and maintain insurance for the benefit of any officer or director in respect of any loss or liability attaching to him or her in respect of any negligence, default, breach of duty or breach of trust, whether or not the Company may otherwise indemnify such officer or director. We will purchase and maintain a directors' and officers' liability policy for such a purpose.

Amendment of Memorandum of Association and Bye-laws

Bermuda law provides that the memorandum of association of a company may be amended by a resolution passed at a general meeting of shareholders. Amendments to most provisions of our bye-laws require an affirmative vote of a majority of our board of directors and a majority of the issued and outstanding shares carrying the right to vote at general meetings at the relevant time. In addition, amendments to certain sections of our bye-laws containing anti-takeover provisions

require an affirmative vote of at least 66% of the directors then in office and at least 66% of the voting power of the issued and outstanding shares carrying the right to vote at general meetings at the relevant time. These provisions make it more difficult for any person to remove or amend any provisions in our bye-laws that may have an anti-takeover effect.

Under Bermuda law, the holders of an aggregate of not less than 20% in par value of the Company's issued share capital or any class thereof have the right to apply to the Supreme Court of Bermuda for an annulment of any amendment of the memorandum of association adopted by shareholders at any general meeting, other than an amendment which alters or reduces a company's share capital as provided in the Companies Act. Where such an application is made, the amendment becomes effective only to the extent that it is confirmed by the Bermuda court. An application for an annulment of an amendment of the memorandum of association must be made within 21 days after the date on which the resolution altering the Company's memorandum of association is passed and may be made on behalf of persons entitled to make the application by one or more of their number as they may appoint in writing for the purpose. No application may be made by shareholders voting in favor of the amendment.

Amalgamations and Mergers

The amalgamation or merger of a Bermuda company with another company or corporation (other than certain affiliated companies) requires the amalgamation or merger to be approved by the company's board of directors and by its shareholders. Unless the company's bye-laws provide otherwise, the approval of 75% of the shareholders voting at such meeting is required to approve the amalgamation or merger, and the quorum for such meeting must be two or more persons holding or representing more than one-third of the issued shares of the company. Our amended and restated bye-laws provide that the approval of the affirmative vote of a majority of votes cast at a meeting to approve the amalgamation or merger shall be sufficient, and the quorum for such meeting shall be two or more persons holding or representing a majority of the voting power of the issued and outstanding voting shares.

Under Bermuda law, in the event of an amalgamation or merger of a Bermuda company with another company or corporation, a shareholder of the Bermuda company who did not vote in favor of the amalgamation or merger and who is not satisfied that fair value has been offered for such shareholder's shares may, within one month of notice of the shareholders meeting, apply to the Supreme Court of Bermuda to appraise the fair value of those shares.

Business Combinations

Although the Companies Act does not contain specific provisions regarding "business combinations" between companies organized under the laws of Bermuda and "interested shareholders," we have included these provisions in our bye-laws. Specifically, our bye-laws contain provisions which prohibit us from engaging in a business combination with an interested shareholder for a period of three years after the date of the transaction in which the person became an interested shareholder, unless, in addition to any other approval that may be required by applicable law:

- prior to the date of the transaction that resulted in the shareholder becoming an interested shareholder, our board of directors approved either the business combination or the transaction that resulted in the shareholder becoming an interested shareholder;

- upon consummation of the transaction that resulted in the shareholder becoming an interested shareholder, the interested shareholder owned at least 85% of the voting power of our issued and outstanding voting shares at the time the transaction commenced; or

- after the date of the transaction that resulted in the shareholder becoming an interested shareholder, the business combination is approved by our board of directors and authorized at an annual or special general meeting of shareholders by the affirmative vote of at least 66^{2}/3% of the voting power of our issued and outstanding voting shares that are not owned by the interested shareholder.

For purposes of these provisions, a "business combination" includes recapitalizations, mergers, amalgamations, consolidations, exchanges, asset sales, leases, certain issues or transfers of shares or other securities and other transactions resulting in a financial benefit to the interested shareholder. An "interested shareholder" is any person or entity that beneficially owns 15% or more of our issued and outstanding voting shares and any person or entity affiliated with or controlling or controlled by that person or entity.

Shareholder Suits

Class actions and derivative actions are generally not available to shareholders under Bermuda law. The Bermuda courts, however, would ordinarily be expected to permit a shareholder to commence an action in the name of a company to remedy a wrong to the company where the act complained of is alleged to be beyond the corporate power of the company or illegal, or would result in the violation of the company's memorandum of association or bye-laws. Furthermore, consideration would be given by a Bermuda court to acts that are alleged to constitute a fraud against the minority shareholders or, for instance, where an act requires the approval of a greater percentage of the company's shareholders than that which actually approved it.

When the affairs of a company are being conducted in a manner that is oppressive or prejudicial to the interests of some part of the shareholders, one or more shareholders may apply to the Supreme Court of Bermuda, which may make such order as it sees fit, including an order regulating the conduct of the company's affairs in the future or ordering the purchase of the shares of any shareholders by other shareholders or by the company.

Our amended and restated bye-laws contain a provision by virtue of which our shareholders waive any claim or right of action that they have, both individually and on our behalf, against any director or officer in relation to any action or failure to take action by such director or officer, except in respect of any fraud or dishonesty of such director or officer. We have been advised by the SEC that in the opinion of the SEC, the operation of this provision as a waiver of the right to sue for violations of federal securities laws would likely be unenforceable in U.S. courts.

Capitalization of Profits and Reserves

Pursuant to our amended and restated bye-laws, our board of directors may (1) capitalize any part of the amount of our share premium or other reserve accounts or any amount credited to our profit and loss account or otherwise available for distribution by applying such sum in paying up unissued shares to be allotted as fully paid bonus shares pro rata (except in connection with the conversion of shares) to the shareholders; or (2) capitalize any sum standing to the credit of a reserve account or sums otherwise available for dividend or distribution by paying up in full, partly paid or nil paid shares of those shareholders who would have been entitled to such sums if they were distributed by way of dividend or distribution.

Untraced Shareholders

Our amended and restated bye-laws provide that our board of directors may forfeit any dividend or other monies payable in respect of any shares that remain unclaimed for six years from the date when such monies became due for payment. In addition, we are entitled to cease sending dividend warrants and checks by post or otherwise to a shareholder if such instruments have been

returned undelivered to, or left uncashed by, such shareholder on at least two consecutive occasions or, following one such occasion, reasonable enquires have failed to establish the shareholder's new address. This entitlement ceases if the shareholder claims a dividend or cashes a dividend check or a warrant.

Certain Provisions of Bermuda Law

We have been designated by the Bermuda Monetary Authority as a non-resident for Bermuda exchange control purposes. This designation allows us to engage in transactions in currencies other than the Bermudan dollar, and there are no restrictions on our ability to transfer funds (other than funds denominated in Bermudan dollars) in and out of Bermuda or to pay dividends to U.S. residents who are holders of our common shares.

The Bermuda Monetary Authority has given its consent for the issue and free transferability of all of the common shares that are the subject of this offering to and between residents and non-residents of Bermuda for exchange control purposes, provided our shares remain listed on an appointed stock exchange, which includes the Nasdaq. Approvals or permissions given by the Bermuda Monetary Authority do not constitute a guarantee by the Bermuda Monetary Authority as to our performance or our creditworthiness. Accordingly, in giving such consent or permissions, neither the Bermuda Monetary Authority nor the Registrar of Companies in Bermuda shall be liable for the financial soundness, performance or default of our business or for the correctness of any opinions or statements expressed in this prospectus. Certain issues and transfers of common shares involving persons deemed resident in Bermuda for exchange control purposes require the specific consent of the Bermuda Monetary Authority. We have sought and have obtained a specific permission from the Bermuda Monetary Authority for the issue and transfer of our common shares up to the amount of our designated capital from time to time, and options, warrants, depository receipts, rights, loan notes, debt instruments and our other securities to persons resident and non-resident for exchange control purposes with the need for prior approval of such issue or transfer.

In accordance with Bermuda law, share certificates are only issued in the names of companies, partnerships or individuals. In the case of a shareholder acting in a special capacity (for example as a trustee), certificates may, at the request of the shareholder, record the capacity in which the shareholder is acting. Notwithstanding such recording of any special capacity, we are not bound to investigate or see to the execution of any such trust.

Registration Rights

Upon the closing of this offering, holders of 35,670,093 Class A common shares (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares), which we refer to as registrable securities, or their transferees will be entitled to the following rights with respect to the registration of such shares for public resale under the Securities Act pursuant to an amended and restated investors rights agreement by and among us and certain of our shareholders, until such shares can otherwise be sold without restriction under Rule 144, or until the rights otherwise terminate pursuant to the terms of the investors' rights agreement. The registration of our common shares as a result of the following rights being exercised would enable holders to trade these shares without restriction under the Securities Act when the applicable registration statement is declared effective.

If at any time beginning 180 days after the closing date of this offering the holders of a majority of the registrable securities request in writing that we effect a registration with respect to all or part of such registrable securities then outstanding, we may be required to register their shares. We are obligated to effect at most one registration in response to these demand registration rights.

If the holders requesting registration intend to distribute their shares by means of an underwriting, the managing underwriter of such offering will have the right to limit the numbers of shares to be underwritten for reasons related to the marketing of the shares.

Piggyback Registration Rights

If at any time after this offering we propose to register any of our Class A common shares under the Securities Act, subject to certain exceptions, the holders of registrable securities will be entitled to notice of the registration and to include their shares of registrable securities in the registration. If our proposed registration involves an underwriting, the managing underwriter of such offering will have the right to limit the number of shares to be underwritten for reasons related to the marketing of the shares.

Form S-3 Registration Rights

If, at any time after we become entitled under the Securities Act to register our shares on a registration statement on Form S-3, 25% of the holders of the registrable securities then outstanding on an as-converted into Class A common shares basis request in writing that we effect a registration with respect to registrable securities at an aggregate price to the public in the offering of at least $5.0 million, we will be required to effect such registration within 20 days after the date of such request; provided, however, that we will not be required to effect such a registration if, within any twelve-month period, we have already effected two registrations on Form S-3 for the holders of registrable securities. On the day we are eligible to use a Form S-3 registration statement, we are obligated to register any then outstanding registrable securities held by affiliates.

Expenses

Ordinarily, other than underwriting discounts and commissions, we will be required to pay all expenses incurred by us related to any registration effected pursuant to the exercise of these registration rights. These expenses may include all registration and filing fees, printing expenses, fees and disbursements of our counsel, reasonable fees and disbursements of a counsel for the selling securityholders and blue-sky fees and expenses.

Termination of Registration Rights

The registration rights terminate upon the earlier of the closing of a deemed liquidation event, as defined in our bye-laws, or, with respect to the registration rights of an individual holder, when the holder can sell all of such holder's registrable securities in a 90-day period without restriction under Rule 144 under the Securities Act.

Certain Corporate Anti-Takeover Provisions

Certain provisions in our amended and restated bye-laws may be deemed to have an anti-takeover effect and may delay, deter or prevent a tender offer or takeover attempt that a shareholder might consider to be in its best interests, including attempts that might result in a premium being paid over the market price for the Class A common shares. These provisions are also designed, in part, to encourage persons seeking to acquire control of us to first negotiate with our board of directors.

Preferred Shares

Pursuant to our amended and restated bye-laws, preferred shares may be issued from time to time, and the board of directors is authorized to determine the rights, preferences, powers, qualifications, limitations and restrictions.

Classified Board

Upon consummation of this offering, in accordance with the terms of our bye-laws, our board of directors will be divided into three classes, class I, class II and class III, with members of each class serving staggered three-year terms after their initial terms of office following their election at the first general meeting of shareholders following this offering. Our amended and restated bye-laws further provide that the authorized number of directors may be changed only by resolution of the board of directors. Any additional directorships resulting from an increase in the number of directors will be distributed among the three classes so that, as nearly as possible, each class will consist of one-third of the directors. Our classified board of directors could have the effect of delaying or discouraging an acquisition of us or a change in our management.

Removal of Directors

Upon consummation of this offering, in accordance with the terms of our amended and restated bye-laws, our directors may be removed only for cause by the affirmative vote of a majority of the votes entitled to be cast by our shareholders entitled to vote at an annual general election of directors. Any vacancy on our board, including a vacancy resulting from an enlargement of our board or from removal for cause not filled by the shareholders at the time, may be filled only by vote of a majority of our directors then in office.

Advance Notice Requirements for Shareholder Proposals and Director Nominations

Our amended and restated bye-laws provide that shareholders seeking to nominate candidates for election as directors or to bring business before an annual meeting of shareholders must provide timely notice of their proposal. Generally, to be timely, a shareholder's notice must be received at our principal executive offices not less than 90 days or more than 120 days prior to the first anniversary date of the last annual general meeting. Our amended and restated bye-laws also specify requirements as to the form and content of a shareholder's notice. These provisions may impede shareholders' ability to bring matters before an annual meeting of shareholders or make nominations for directors at an annual meeting of shareholders.

Choice of Jurisdiction

Our amended and restated by-laws provide that, unless we consent in writing to the selection of an alternative jurisdiction, any dispute that arises concerning the Companies Act or out of or in connection with our bye-laws, including any question regarding the existence and scope of any bye-law and/or whether there has been a breach of the Companies Act or the bye-laws by any of our officers or directors (whether or not such a claim is brought in the name of a shareholder or in the name of our company) shall be subject to the jurisdiction of the Supreme Court of Bermuda.

Amendment of Certain Bye-Laws

Amendments to certain sections of our amended and restated bye-laws containing anti-takeover provisions will require an affirmative vote of at least 66% of the directors and at least 66% of the voting power of the issued and outstanding shares.

Registrar and Transfer Agent

A register of holders of the Class A common shares will be maintained by Conyers Corporate Services (Bermuda) Limited in Bermuda, and a branch register will be maintained in the United States by American Stock Transfer & Trust Company LLC, which will also serve as transfer agent. The transfer agent's address is 6201 15th Avenue, Brooklyn, NY 11219.

Listing

We have applied to have our Class A common shares listed on The Nasdaq Global Market under the symbol "KNSA."

SHARES ELIGIBLE FOR FUTURE SALE

Prior to this offering, there has been no public market for our Class A common shares. Future sales of substantial amounts of Class A common shares in the public market after this offering, or the perception that such sales may occur, could adversely affect the market price of our Class A common shares and could impair our future ability to raise equity capital.

Upon the closing of this offering, we will have outstanding an aggregate of 13,270,569 Class A common shares, 12,995,954 Class A1 common shares, 4,638,855 Class B common shares and 16,057,618 Class B1 common shares, assuming the issuance of 7,000,000 Class A common shares offered by us in this offering, the conversion of all our outstanding preferred shares into 5,546,019 Class A common shares, 12,995,954 Class A1 common shares, 1,070,502 Class B common shares and 16,057,618 Class B1 common shares upon the closing of this offering, and no exercise of options after March 31, 2018. Of these shares, all Class A common shares sold in this offering will be freely tradable without restriction or further registration under the Securities Act, except for any shares purchased by our "affiliates," as that term is defined in Rule 144 under the Securities Act, whose sales would be subject to the Rule 144 resale restrictions described below, other than the holding period requirement.

The aggregate remaining 39,962,996 Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares will be "restricted securities," as that term is defined in Rule 144 under the Securities Act. These restricted securities are eligible for public sale only if they are registered under the Securities Act or if they qualify for an exemption from registration under Rules 144 or 701 under the Securities Act, which are summarized below. We expect that all of these shares will be subject to the 180-day lock-up period under the lock-up agreements described below. Upon expiration of the lock-up period, we estimate that 39,837,144 Class A common shares will be available for sale in the public market (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares), subject in some cases to applicable volume limitations under Rule 144.

In addition, of the 4,702,190 Class A common shares that were subject to share options outstanding as of March 31, 2018, options to purchase 955,119 Class A common shares were vested as of March 31, 2018 and, upon exercise, these shares will be eligible for sale subject to the lock-up agreements described below and Rules 144 and 701 under the Securities Act.

Lock-Up Agreements

We and all of our equity holders as of the date of this prospectus, including each of our executive officers and directors, have entered into or will enter into lock-up agreements with the underwriters or otherwise agree, subject to certain exceptions, that we and they will not, directly or indirectly, offer, sell, contract to sell, pledge, grant any option to purchase, make any short sale or otherwise dispose of or hedge any of our common shares, any options or warrants to purchase our common shares, or any securities convertible into, or exchangeable for or that represent the right to receive our common shares, without the prior written consent of Goldman Sachs & Co. LLC and J.P. Morgan Securities LLC for a period of 180 days from the date of this prospectus.

Rule 144

Affiliate Resales of Restricted Securities

In general, beginning 90 days after the effective date of the registration statement of which this prospectus is a part, a person who is an affiliate of ours, or who was an affiliate at any time during the 90 days before a sale, who has beneficially owned our Class A common shares for at least six

months would be entitled to sell in "broker's transactions" or certain "riskless principal transactions" or to market makers, a number of shares within any three-month period that does not exceed the greater of:

- 1% of the number of our Class A common shares then outstanding, which will equal approximately 132,705 shares immediately after this offering; or
- the average weekly trading volume in our Class A common shares on The Nasdaq Global Market during the four calendar weeks preceding the filing of a notice on Form 144 with respect to such sale.

Affiliate resales under Rule 144 are also subject to the availability of current public information about us. In addition, if the number of shares being sold under Rule 144 by an affiliate during any three-month period exceeds 5,000 shares or has an aggregate sale price in excess of $50,000, the seller must file a notice on Form 144 with the SEC and Nasdaq concurrently with either the placing of a sale order with the broker or the execution directly with a market maker.

Non-Affiliate Resales of Restricted Securities

In general, beginning 90 days after the effective date of the registration statement of which this prospectus is a part, a person who is not an affiliate of ours at the time of sale, and has not been an affiliate at any time during the three months preceding a sale, and who has beneficially owned our Class A common shares for at least six months but less than a year, is entitled to sell such shares subject only to the availability of current public information about us. If such person has held our shares for at least one year, such person can resell under Rule 144(b)(1) without regard to any Rule 144 restrictions, including the 90-day public company requirement and the current public information requirement.

Non-affiliate resales are not subject to the manner of sale, volume limitation or notice filing provisions of Rule 144.

Rule 701

In general, under Rule 701, any of an issuer's employees, directors, officers, consultants or advisors who purchases shares from the issuer in connection with a compensatory share or option plan or other written agreement before the effective date of a registration statement under the Securities Act is entitled to sell such shares 90 days after such effective date in reliance on Rule 144. An affiliate of the issuer can resell shares in reliance on Rule 144 without having to comply with the holding period requirement, and non-affiliates of the issuer can resell shares in reliance on Rule 144 without having to comply with the current public information and holding period requirements.

The SEC has indicated that Rule 701 will apply to typical share options granted by an issuer before it becomes subject to the reporting requirements of the Exchange Act, along with the shares acquired upon exercise of such options, including exercises after an issuer becomes subject to the reporting requirements of the Exchange Act.

Equity Plans

We intend to file one or more registration statements on Form S-8 under the Securities Act to register all Class A common shares subject to outstanding share options and Class A common shares issued or issuable under our share plans. We expect to file the registration statement covering shares offered pursuant to our share plans shortly after the date of this prospectus, permitting the resale of such shares by non-affiliates in the public market without restriction under

the Securities Act and the sale by affiliates in the public market, subject to compliance with the resale provisions of Rule 144.

Registration Rights

Upon the closing of this offering, the holders of 35,670,093 Class A common shares (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares), or their transferees, will be entitled to various rights with respect to the registration under the Securities Act of these shares and any common shares issued as a dividend or other distribution with respect to these Class A common shares. Registration of these shares under the Securities Act would result in these shares becoming fully tradable without restriction under the Securities Act immediately upon the effectiveness of the registration statement, except for shares purchased by affiliates. See "Description of Share Capital — Registration Rights" for additional information.

BERMUDA COMPANY CONSIDERATIONS

Our corporate affairs will be governed by our memorandum of association and our amended and restated bye-laws that will become effective immediately following the closing of this offering and by the corporate law of Bermuda. The provisions of the Companies Act, which applies to us, differ in certain material respects from laws generally applicable to U.S. companies incorporated in the State of Delaware and their stockholders. The following is a summary of significant differences between the Companies Act and Bermuda common law applicable to us and our shareholders and the provisions of the Delaware General Corporation Law applicable to U.S. companies organized under the laws of Delaware and their stockholders.

Bermuda	Delaware
Shareholder Meetings	
• May be called by the board of directors and must be called upon the request of shareholders holding not less than 10% of the paid-up capital of the company carrying the right to vote at general meetings.	• May be held at such time or place as designated in the certificate of incorporation or the bylaws, or if not so designated, as determined by the board of directors.
• May be held in or outside Bermuda.	• May be held in or outside of Delaware.
• Notice:	• Notice:
◦ Shareholders must be given at least five days' advance notice of a general meeting, but the unintentional failure to give notice to any person does not invalidate the proceedings at a meeting.	◦ Written notice shall be given not less than ten nor more than 60 days before the meeting.
◦ Notice of general meetings must specify the place, the day and hour of the meeting and in the case of special general meetings, the general nature of the business to be considered.	◦ Whenever stockholders are required to take any action at a meeting, a written notice of the meeting shall be given which shall state the place, if any, date and hour of the meeting, the means of remote communication, if any, the record date for determining the stockholders entitled to vote at the meeting if such date is different from the record date for determining stockholders entitled to notice, and in the case of a special meeting, the purpose for which the meeting is called.

Bermuda	Delaware
Shareholders' Voting Rights	
• Shareholders may act by written consent to elect directors. Shareholders may not act by written consent to remove a director or auditor.	• With limited exceptions, stockholders may act by written consent to elect directors unless prohibited by the certificate of incorporation.
• Generally, except as otherwise provided in the bye-laws, or the Companies Act, any action or resolution requiring approval of the shareholders may be passed by a simple majority of votes cast. Any person authorized to vote may authorize another person or persons to act for him or her by proxy.	• Any person authorized to vote may authorize another person or persons to act for him or her by proxy.
• The voting rights of shareholders are regulated by a company's bye-laws and, in certain circumstances, by the Companies Act. The bye-laws may specify the number to constitute a quorum and if the bye-laws permit, a general meeting of the shareholders of a company may be held with only one individual present if the requirement for a quorum is satisfied.	• For stock corporations, the certificate of incorporation or bylaws may specify the number to constitute a quorum, but in no event shall a quorum consist of less than one-third of shares entitled to vote at a meeting. In the absence of such specifications, a majority of shares entitled to vote shall constitute a quorum.
	• When a quorum is once present to organize a meeting, it is not broken by the subsequent withdrawal of any stockholders.
• The bye-laws may provide for cumulative voting.	• The certificate of incorporation may provide for cumulative voting.

Bermuda	Delaware
Mergers or Sale of Assets	
• The amalgamation or merger of a Bermuda company with another company or corporation (other than certain affiliated companies) requires the amalgamation or merger agreement to be approved by the company's board of directors and by its shareholders. Unless the company's bye-laws provide otherwise, the approval of 75% of the shareholders voting at such meeting is required to approve the amalgamation or merger agreement, and the quorum for such meeting must be two or more persons holding or representing more than one-third of the issued shares of the company.	• Any two or more corporations existing under the laws of the state may merge into a single corporation pursuant to a board resolution and upon the majority vote by stockholders of each constituent corporation at an annual or special meeting.
• Every company may at any meeting of its board of directors sell, lease or exchange all or substantially all of its property and assets as its board of directors deems expedient and in the best interests of the company to do so when authorized by a resolution adopted by the holders of a majority of issued and outstanding shares of a company entitled to vote.	• Every corporation may at any meeting of the board sell, lease or exchange all or substantially all of its property and assets as its board deems expedient and for the best interests of the corporation when so authorized by a resolution adopted by the holders of a majority of the outstanding stock of a corporation entitled to vote.
• Any company that is the wholly owned subsidiary of a holding company, or one or more companies which are wholly owned subsidiaries of the same holding company, may amalgamate or merge without the vote or consent of shareholders provided that the approval of the board of directors is obtained and that a director or officer of each such company signs a statutory solvency declaration in respect of the relevant company.	• Any corporation owning at least 90% of the outstanding shares of each class of another corporation may merge the other corporation into itself and assume all of its obligations without the vote or consent of stockholders if one of the two corporations is a Delaware entity and the laws or a foreign corporation do not prohibit such merger. However, in a case where the parent corporation is not the surviving corporation, the proposed merger shall be approved by a majority of the outstanding stock of the parent corporation entitled to vote at a duly called stockholder meeting.
• Any mortgage, charge or pledge of a company's property and assets may be authorized without the consent of shareholders subject to any restrictions under the bye-laws.	• Any mortgage or pledge of a corporation's property and assets may be authorized without the vote or consent of stockholders, except to the extent that the certificate of incorporation otherwise provides.

Bermuda	Delaware
Directors	

• The board of directors must consist of at least one director.
• The number of directors is fixed by the bye-laws, and any changes to such number must be approved by the board of directors and/or the shareholders in accordance with the company's bye-laws.
	• The board of directors must consist of at least one member.
	• Number of board members shall be fixed by the bylaws, unless the certificate of incorporation fixes the number of directors, in which case a change in the number shall be made only by amendment of the certificate of incorporation.
	• Removal:
	  • Any or all of the directors may be removed, with or without cause, by the holders of a majority of the shares entitled to vote unless the certificate of incorporation otherwise provides.
	  • In the case of a classified board, stockholders may effect removal of any or all directors only for cause.
	  • In the case of a corporation with cumulative voting, if less than the entire board is to be removed, no director may be removed without cause if the votes against removal would be sufficient to elect such director using cumulative voting.

Bermuda	Delaware
Duties of Directors	

• The Companies Act authorizes the directors of a company, subject to its bye-laws, to exercise all powers of the company except those that are required by the Companies Act or the company's bye-laws to be exercised by the shareholders of the company. At common law, members of a board of directors owe a fiduciary duty to the company to act in good faith in their dealings with or on behalf of the company and exercise their powers and fulfill the duties of their office honestly. This duty includes the following essential elements:
  ◦ a duty to act in good faith in the best interests of the company;
  ◦ a duty not to make a personal profit from opportunities that arise from the office of director;
  ◦ a duty to avoid conflicts of interest; and
  ◦ a duty to exercise powers for the purpose for which such powers were intended.
• The Companies Act imposes a duty on directors and officers of a Bermuda company:
  ◦ to act honestly and in good faith with a view to the best interests of the company; and
  ◦ to exercise the care, diligence and skill that a reasonably prudent person would exercise in comparable circumstances.
• The Companies Act also imposes various duties on directors and officers of a company with respect to certain matters of management and administration of the company. Under Bermuda law, directors and officers generally owe fiduciary duties to the company itself, not to the company's individual shareholders, creditors or any class thereof.
	• Under Delaware law, the business and affairs of a corporation are managed by or under the direction of its board of directors except as may be otherwise provided in its certificate of incorporation. In exercising their powers, directors are charged with a fiduciary duty of care to protect the interests of the corporation and a fiduciary duty of loyalty to act in the best interests of its stockholders. The duty of care requires that a director act in good faith, with the care that an ordinarily prudent person would exercise under similar circumstances. Under this duty, a director must inform himself of, and disclose to stockholders, all material information reasonably available regarding a significant transaction. The duty of loyalty requires that a director act in a manner he reasonably believes to be in the best interests of the corporation. He must not use his corporate position for personal gain or advantage. This duty prohibits self-dealing by a director and mandates that the best interest of the corporation and its stockholders take precedence over any interest possessed by a director, officer or controlling shareholder and not shared by the stockholders generally.
	• In general, actions of a director are presumed to have been made on an informed basis, in good faith and in the honest belief that the action taken was in the best interests of the corporation. However, this presumption may be rebutted by evidence of a breach of one of the fiduciary duties. Should such evidence be presented concerning a transaction by a director, a director must prove the procedural fairness of the transaction, and that the transaction was of fair value to the corporation.

Bermuda	Delaware
Takeovers	

• An acquiring party is generally able to acquire compulsorily the common shares of minority holders of a company in the following ways:
  • By a procedure under the Companies Act known as a "scheme of arrangement." A scheme of arrangement could be effected by obtaining the agreement of the company and of holders of common shares, representing in the aggregate a majority in number and at least 75% in value of the common shareholders present and voting at a court ordered meeting held to consider the scheme of arrangement. The scheme of arrangement must then be sanctioned by the Bermuda Supreme Court. If a scheme of arrangement receives all necessary agreements and sanctions, upon the filing of the court order with the Registrar of Companies in Bermuda, all holders of common shares could be compelled to sell their shares under the terms of the scheme of arrangement.
  • By acquiring pursuant to a tender offer 90% of the shares or class of shares not already owned by, or by a nominee for, the acquiring party (the offeror), or any of its subsidiaries. If an offeror has, within four months after the making of an offer for all the shares or class of shares not owned by, or by a nominee for, the offeror, or any of its subsidiaries, obtained the approval of the holders of 90% or more of all the shares to which the offer relates, the offeror may, at any time within two months beginning with the date on which the approval was obtained, by notice compulsorily acquire the shares of any nontendering shareholder on the same terms as the original offer unless the Supreme Court of Bermuda (on application made within a one-month period from the date of the offeror's notice of its intention to acquire such shares) orders otherwise.
	• Delaware law provides that a parent corporation, by resolution of its board of directors and without any stockholder vote, may merge with any subsidiary of which it owns at least 90% of each class of its capital stock. Upon any such merger, and in the event the parent corporate does not own all of the stock of the subsidiary, dissenting stockholders of the subsidiary are entitled to certain appraisal rights.
	• Delaware law also provides, subject to certain exceptions, that if a person acquires 15% of voting stock of a company or is an affiliate or associate of the corporation and owned 15% of voting stock within a 3-year period, the person is an "interested stockholder" and may not engage in "business combinations" with the company for a period of three years from the time the person acquired 15% or more of voting stock.

Bermuda	Delaware
• Where the acquiring party or parties hold not less than 95% of the shares or a class of shares of the company, by acquiring, pursuant to a notice given to the remaining shareholders or class of shareholders, the shares of such remaining shareholders or class of shareholders. When this notice is given, the acquiring party is entitled and bound to acquire the shares of the remaining shareholders on the terms set out in the notice, unless a remaining shareholder, within one month of receiving such notice, applies to the Supreme Court of Bermuda for an appraisal of the value of their shares. This provision only applies where the acquiring party offers the same terms to all holders of shares whose shares are being acquired.	
Dissenter's Rights of Appraisal	
• A dissenting shareholder (that did not vote in favor of the amalgamation or merger) of a Bermuda exempted company is entitled to be paid the fair value of his or her shares in an amalgamation or merger.	• With limited exceptions, appraisal rights shall be available for the shares of any class or series of stock of a corporation in a merger or consolidation. • The certificate of incorporation may provide that appraisal rights are available for shares as a result of an amendment to the certificate of incorporation, any merger or consolidation or the sale of all or substantially all of the assets.

Bermuda	Delaware
Dissolution	
• Under Bermuda law, a solvent company may be wound up by way of a shareholders' voluntary liquidation. Prior to the company entering liquidation, a majority of the directors shall each make a statutory declaration, which states that the directors have made a full enquiry into the affairs of the company and have formed the opinion that the company will be able to pay its debts within a period of 12 months of the commencement of the winding up and must file the statutory declaration with the Registrar of Companies in Bermuda. The general meeting will be convened primarily for the purposes of passing a resolution that the company be wound up voluntarily and appointing a liquidator. The winding up of the company is deemed to commence at the time of the passing of the resolution.	• Under Delaware law, a corporation may voluntarily dissolve (1) if a majority of the board of directors adopts a resolution to that effect and the holders of a majority of the issued and outstanding shares entitled to vote thereon vote for such dissolution; or (2) if all stockholders entitled to vote thereon consent in writing to such dissolution.
Shareholders' Derivative Actions	
• Class actions and derivative actions are generally not available to shareholders under Bermuda law. Bermuda courts, however, would ordinarily be expected to permit a shareholder to commence an action in the name of a company to remedy a wrong to the company where the act complained of is alleged to be beyond the corporate power of the company or illegal, or would result in the violation of the company's memorandum of association or bye-laws. Furthermore, consideration would be given by a Bermuda court to acts that are alleged to constitute a fraud against the minority shareholders or, for instance, where an act requires the approval of a greater percentage of the company's shareholders than that which actually approved it.	• In any derivative suit instituted by a stockholder of a corporation, it shall be averred in the complaint that the plaintiff was a stockholder of the corporation at the time of the transaction of which such stockholder complains or that such stockholder's stock thereafter devolved upon the stockholder by operation of law.

MATERIAL BERMUDA AND U.S. FEDERAL INCOME TAX CONSIDERATIONS

The following is a discussion of the material Bermuda and U.S. federal income tax considerations that may be relevant to an investment decision by a potential investor with respect to our common shares.

Bermuda Tax Considerations

Taxation of the Company

Under current Bermuda law, there is no income, corporate or profits tax or withholding tax, capital gains tax or capital transfer tax, estate or inheritance tax payable by us or our shareholders, other than shareholders ordinarily resident in Bermuda, if any. The Company has received from the Minister of Finance under The Exempted Undertaking Tax Protection Act 1966, as amended, an assurance that, in the event that Bermuda enacts legislation imposing tax computed on profits, income, any capital asset, gain or appreciation, or any tax in the nature of estate duty or inheritance, then the imposition of any such tax will not be applicable to the Company or to any of their operations or their shares, debentures or other obligations, until March 31, 2035. This assurance is subject to the proviso that it is not to be construed so as to prevent the application of any tax or duty to such persons as are ordinarily resident in Bermuda or to prevent the application of any tax payable in accordance with the provisions of the Land Tax Act 1967 or otherwise payable in relation to any property leased to the Company. The Company pays annual Bermuda government fees which fees are calculated on a sliding scale based on the assessable capital of the company. In addition, all entities employing individuals in Bermuda are required to pay a payroll tax and there are other sundry taxes payable, directly or indirectly, to the Bermuda government.

Taxation of Shareholders

Currently, there is no Bermuda income, corporate or profits tax or withholding tax, capital gains tax or capital transfer tax, estate or inheritance tax payable by holders of our shares, other than shareholders ordinarily resident in Bermuda, if any.

Material U.S. Federal Income Tax Considerations to U.S. Holders

The following discussion describes the material U.S. federal income tax consequences to U.S. Holders (as defined below) under present law of an investment in the Class A common shares. The effects of any applicable state or local laws, or other U.S. federal tax laws such as estate and gift tax laws, or the alternative minimum tax or the Medicare contribution tax on net investment income, are not discussed. This summary applies only to investors who acquire the Class A common shares in exchange for cash, hold the Class A common shares as capital assets (generally, property held for investment) and who have the U.S. dollar as their functional currency. This discussion is based on the U.S. Internal Revenue Code of 1986, as amended, or the Code, U.S. Treasury regulations promulgated thereunder, judicial decisions, published rulings and administrative pronouncements of the U.S. Internal Revenue Service, or the IRS, all as in effect as of the date of this prospectus. All of the foregoing authorities are subject to change, which change could apply retroactively and could affect the tax consequences described below.

The following discussion does not address all U.S. federal income tax consequences relevant to a holder's particular circumstances or to holders subject to particular rules, including:

- U.S. expatriates and certain former citizens or long-term residents of the United States;
- persons whose functional currency is not the U.S. dollar;

- persons holding Class A common shares as part of a hedge, straddle or other risk reduction strategy or as part of a conversion transaction or other integrated investment;
- banks, insurance companies and other financial institutions;
- real estate investment trusts or regulated investment companies;
- brokers, dealers or traders in securities, commodities or currencies;
- S corporations or entities or arrangements treated as partnerships for U.S. federal income tax purposes;
- tax-exempt organizations or governmental organizations;
- individual retirement accounts or other tax deferred accounts;
- persons who acquired the Class A common shares pursuant to the exercise of any employee share option or otherwise as compensation;
- persons that own or are deemed to own 10% or more of our stock by vote or value;
- persons subject to special tax accounting rules as a result of any item of gross income with respect to the Class A common shares being taken into account in an applicable financial statement;
- persons that hold Class A common shares through a permanent establishment or fixed base outside the United States; and
- persons deemed to sell Class A common shares under the constructive sale provisions of the Code.

We believe we are a "controlled foreign corporation" for U.S. federal income tax purposes, and therefore, if you are a U.S. shareholder owning 10% or more of our stock by vote or value directly, indirectly or constructively, the U.S. federal income tax consequences to you of owning our Class A common shares may be significantly different than those described below. If you own 10% or more of our stock by vote or value directly, indirectly or constructively, you should consult your tax advisors regarding the U.S. federal income tax consequence of your investment in our Class A common shares.

U.S. HOLDERS ARE URGED TO CONSULT THEIR TAX ADVISORS REGARDING THE APPLICATION OF THE U.S. FEDERAL TAX RULES TO THEIR PARTICULAR CIRCUMSTANCES AS WELL AS THE U.S. STATE AND LOCAL AND NON-U.S. TAX CONSEQUENCES TO THEM OF THE PURCHASE, OWNERSHIP AND DISPOSITION OF CLASS A COMMON SHARES.

For purposes of this discussion, a "U.S. Holder" is a beneficial owner of Class A common shares that, for U.S. federal income tax purposes, is or is treated as any of the following:

- an individual who is a citizen or resident of the United States;
- a corporation (or another entity taxable as a corporation) created or organized under the laws of the United States, any state thereof, or the District of Columbia;
- an estate, the income of which is subject to U.S. federal income tax regardless of its source; or
- a trust that (1) is subject to the supervision of a U.S. court and the control of one or more "United States persons" (within the meaning of Section 7701(a)(30) of the Code), or (2) has a valid election in effect to be treated as a United States person for U.S. federal income tax purposes.

If you are an entity taxable as a partnership for U.S. federal income tax purposes that holds Class A common shares, your tax treatment generally will depend on your status and the activities of the partnership. Partnerships holding Class A common shares and the partners in such partnerships should consult their tax advisors regarding the U.S. federal income tax consequences applicable to them.

Taxation of Dividends and Other Distributions on the Class A Common Shares

The discussion in this section "Taxation of Dividends and Other Distributions on the Class A Common Shares" is subject to the discussion regarding passive foreign investment companies below. As discussed above under "Dividend Policy," the Company does not currently intend to declare dividends on the Class A common shares in the foreseeable future. In the event the Company does pay dividends, the gross amount of any distribution to you with respect to the Class A common shares will be included in your gross income as dividend income when actually or constructively received to the extent that the distribution is paid out of our current or accumulated earnings and profits (as determined under U.S. federal income tax principles). To the extent the amount of the distribution exceeds our current and accumulated earnings and profits, it will be treated first as a return of your tax basis in the Class A common shares, and to the extent the amount of the distribution exceeds your tax basis, the excess will be taxed as capital gain. We do not intend to calculate our earnings and profits under U.S. federal income tax principles. Therefore, a U.S. Holder should expect that distributions will generally be reported as ordinary dividend income for such purposes. Dividends we pay will not be eligible for the dividends-received deduction available to corporations in respect of dividends received from U.S. corporations.

Subject to certain limitations, dividends paid by qualified foreign corporations to certain non-corporate U.S. Holders may be taxable at preferential tax rates. A non-U.S. corporation is generally treated as a qualified foreign corporation with respect to dividends paid on stock that is readily tradable on a securities market in the United States, such as Nasdaq, on which the Company has applied to list the Class A common shares. However, the preferential tax rates discussed above will not apply if we are treated as a passive foreign investment company with respect to the U.S. Holder for the taxable year in which a dividend is paid or the preceding year. Non-corporate U.S. Holders should consult their tax advisers regarding the availability of the reduced tax rate on dividends. Dividends will be included in a U.S. Holder's income on the date of the U.S. Holder's receipt of the dividend.

Dividends will generally constitute foreign source income for foreign tax credit limitation purposes. Any tax withheld with respect to distributions on the Class A common shares may, subject to a number of complex limitations, be claimed as a foreign tax credit against such U.S. Holder's U.S. federal income tax liability or may be claimed as a deduction for U.S. federal income tax purposes. The limitation on foreign taxes eligible for credit is calculated separately with respect to specific classes of income. For this purpose, dividends distributed by us with respect to the Class A common shares generally will constitute "passive category income." The rules with respect to the foreign tax credit are complex and may depend upon a U.S. Holder's particular circumstances. You should consult your tax advisor regarding the availability of the foreign tax credit under your particular circumstances.

Taxation of Disposition of the Class A Common Shares

The discussion in this section "Taxation of Dispositions of Class A Common Shares" is subject to the discussion regarding passive foreign investment companies below. You will recognize gain or loss on any sale, exchange or other taxable disposition of Class A common shares equal to the difference between the amount realized (in U.S. dollars) on the disposition and your tax basis (in U.S. dollars) in the Class A common shares. Any such gain or loss will be capital gain or loss, and

will be long-term capital gain or loss if you have held the Class A common shares for more than one year at the time of the disposition. Otherwise, such gain or loss will be short-term capital gain or loss. Long-term capital gains recognized by certain non-corporate U.S. Holders, including individuals, generally will be taxable at reduced rates. The deductibility of capital losses is subject to limitations. Any such gain or loss you recognize generally will be treated as U.S. source income or loss for foreign tax credit limitation purposes. You should consult your tax advisor regarding the proper treatment of gain or loss in your particular circumstances.

Passive Foreign Investment Company

Because we do not expect to earn revenue from our business operations during the current taxable year, and because our sole source of income currently is interest on bank accounts, we believe we will likely be a PFIC for our current taxable year. A non-U.S. corporation will be classified as a PFIC for any taxable year in which, after applying certain look-through rules, either:

- at least 75% of its gross income for such taxable year is passive income, or
- at least 50% of the value of its assets (based on an average of the quarterly values of the assets during a taxable year) is attributable to assets that produce or are held for the production of passive income.

For purposes of the above calculations, if a non-U.S. corporation owns, directly or indirectly, 25% or more of the total value of the outstanding shares of another corporation, it will be treated as if it (a) held a proportionate share of the assets of such other corporation and (b) received directly a proportionate share of the income of such other corporation. Passive income generally includes dividends, interest, rents, royalties and capital gains, but generally excludes rents and royalties which are derived in the active conduct of a trade or business and which are received from a person other than a related person.

A separate determination must be made each taxable year as to whether we are a PFIC (after the close of each such taxable year). Because the value of our assets for purposes of the asset test will generally be determined by reference to the market price of the Class A common shares, our PFIC status will depend in large part on the market price of the Class A common shares, which may fluctuate significantly. In addition, changes in the composition of our income or assets may cause us to become a PFIC.

If we are classified as a PFIC in any year with respect to which a U.S. Holder owns Class A common shares, we will continue to be treated as a PFIC with respect to such U.S. Holder in all succeeding years during which the U.S. Holder owns the Class A common shares, regardless of whether we continue to meet the tests described above unless we cease to be a PFIC and the U.S. Holder (1) has made a "deemed sale" election under the PFIC rules, (2) the U.S. Holder has a valid mark-to-market election in effect (as described below) or (3) the U.S. Holder makes a QEF Election (defined below) with respect to all taxable years in which we are a PFIC during such U.S. Holder's holding period in which we are a PFIC or makes a purging election to cause a deemed sale of the PFIC shares at their fair market value in conjunction with a QEF Election (see discussion below regarding such elections). If a U.S. Holder makes a deemed sale election, such U.S. Holder will be deemed to have sold the common shares held by such U.S. Holder at their fair market value, and any gain from such deemed sale would be subject to the rules described below. After the deemed sale election, so long as we do not become a PFIC in a subsequent taxable year, a U.S. Holder's Class A common shares subject to such election will not be treated as shares in a PFIC, and the rules described below with respect to any "excess distributions" or any gain from an actual sale or other disposition of the Class A common shares will not apply. U.S. Holders should consult their tax advisors as to the possibility and consequences of making a deemed sale election if we cease to be a PFIC.

For each taxable year we are treated as a PFIC with respect to you, you will be subject to special tax rules with respect to any "excess distribution" you receive and any gain you realize from a sale or other disposition (including a pledge) of Class A common shares, unless you (i) make a QEF Election (as defined below) with respect to all taxable years of your holding period during which we are a PFIC (as discussed below) or make a purging election to cause a deemed sale of the PFIC shares at their fair market value in conjunction with a QEF Election (see discussion below regarding such elections) or (ii) make a "mark-to-market" election as discussed below. Distributions you receive in a taxable year that are greater than 125% of the average annual distributions you received during the shorter of the three preceding taxable years or your holding period for the Class A common shares will be treated as an excess distribution. Under these special tax rules, if you receive any excess distribution or realize any gain from a sale or other disposition of the Class A common shares:

- the excess distribution or gain will be allocated ratably over your holding period for the Class A common shares,
- the amount allocated to the current taxable year, and any taxable year before the first taxable year in which we were a PFIC, will be treated as ordinary income, and
- the amount allocated to each other year will be subject to the highest tax rate in effect for that year and an interest charge generally applicable to underpayments of tax will be imposed on the resulting tax attributable to each such year.

The tax liability for amounts allocated to years before the year of disposition or "excess distribution" cannot be offset by any net operating losses for such years, and gains (but not losses) realized on the sale of Class A common shares cannot be treated as capital, even if you hold the Class A common shares as capital assets.

If we are treated as a PFIC with respect to you for any taxable year, to the extent any of our subsidiaries are also PFICs, you will be deemed to own your proportionate share of any such lower-tier PFICs, and you may be subject to the rules described in the preceding two paragraphs with respect to the shares of such lower-tier PFICs you would be deemed to own. As a result, you may incur liability for any "excess distribution" described above if we receive a distribution from such lower-tier PFICs or if any shares in such lower-tier PFICs are disposed of (or deemed disposed of). You should consult your tax advisor regarding the application of the PFIC rules to any of our subsidiaries.

A U.S. Holder of "marketable stock" (as defined below) in a PFIC may make a mark-to-market election for such stock to elect out of the general tax treatment for PFICs discussed above. If you make a mark-to-market election for the Class A common shares, you will include in income for each year we are a PFIC an amount equal to the excess, if any, of the fair market value of the Class A common shares as of the close of your taxable year over your adjusted basis in such Class A common shares. Accordingly, a mark-to-market election may accelerate the recognition of income without a corresponding receipt of cash. You are allowed a deduction for the excess, if any, of the adjusted basis of the Class A common shares over their fair market value as of the close of the taxable year. However, deductions are allowable only to the extent of any net mark-to-market gains on the Class A common shares included in your income for prior taxable years. Amounts included in your income under a mark-to-market election, as well as gain on the actual sale or other disposition of Class A common shares, are treated as ordinary income. Ordinary loss treatment also applies to the deductible portion of any mark-to-market loss on Class A common shares, as well as to any loss realized on the actual sale or disposition of Class A common shares to the extent the amount of such loss does not exceed the net mark-to-market gains previously included for the Class A common shares. Your basis in the Class A common shares will be adjusted to reflect any such income or loss amounts. If you make a valid mark-to-market election, the tax rules that apply

to distributions by corporations which are not PFICs would apply to distributions by us, except the lower applicable tax rates for qualified dividend income would not apply. If we cease to be a PFIC when you have a mark-to-market election in effect, gain or loss realized by you on the sale of Class A common shares will be a capital gain or loss and taxed in the manner described above under "Taxation of Disposition of the Class A Common Shares."

The mark-to-market election is available only for "marketable stock," which is stock that is traded in other than de minimis quantities on at least 15 days during each calendar quarter, or regularly traded, on a qualified exchange or other market, as defined in applicable U.S. Treasury regulations. Any trades that have as their principal purpose meeting this requirement will be disregarded. The Class A common shares have been approved for listing on The Nasdaq Global Select Market and, accordingly, provided the Class A common shares are regularly traded, if you are a holder of Class A common shares, the mark-to-market election would be available to you if we are a PFIC. Once made, the election cannot be revoked without the consent of the IRS unless the Class A common shares cease to be marketable stock. If we are a PFIC for any year in which a U.S. Holder owns Class A common shares but before a mark-to-market election is made, the interest charge rules described above will apply to any mark-to market gain recognized in the year the election is made. If any of our subsidiaries are or become PFICs, the mark-to-market election will not be available with respect to shares of such subsidiaries that are treated as owned by you. Consequently, you could be subject to the PFIC rules with respect to income of the lower-tier PFICs the value of which already had been taken into account indirectly via mark-to-market adjustments. You should consult your tax advisor as to the availability and desirability of a mark-to-market election, as well as the impact of such election on interests in any lower-tier PFICs.

Alternatively, a U.S. Holder can make an election, if we provide the necessary information, to treat us and each lower-tier PFIC (if any) as a qualified electing fund (a "QEF Election") in the first taxable year we (and any relevant subsidiaries) are treated as a PFIC with respect to the holder. If such election remains in place while we and any lower-tier PFIC subsidiaries are PFICs, we and our subsidiaries will not be treated as PFICs with respect to such U.S. Holder when we cease to be a PFIC. A U.S. Holder must make the QEF Election for each PFIC by attaching a separate properly completed IRS Form 8621 for each PFIC to the holder's timely filed U.S. federal income tax return. We will provide the information necessary for a U.S. Holder to make a QEF Election with respect to us and will cause each lower-tier PFIC we control to provide such information.

If a U.S. Holder makes a QEF Election with respect to a PFIC, the holder will be currently taxable on its pro rata share of the PFIC's ordinary earnings and net capital gain (at ordinary income and capital gain rates, respectively) for each taxable year that the entity is classified as a PFIC. Accordingly, a QEF election may accelerate the recognition of income without a corresponding receipt of cash. If a U.S. Holder makes a QEF Election with respect to us, any distributions paid by us out of our earnings and profits that were previously included in the holder's income under the QEF Election would not be taxable to the holder. A U.S. Holder will increase its tax basis in its Class A common shares by an amount equal to any income included under the QEF Election and will decrease its tax basis by any amount distributed that is not included in the holder's income. In addition, a U.S. Holder will recognize capital gain or loss on the disposition of Class A common shares in an amount equal to the difference between the amount realized and the holder's adjusted tax basis in the Class A common shares. U.S. Holders should note that if they make QEF Elections with respect to us and any lower-tier PFICs, they may be required to pay U.S. federal income tax with respect to their Class A common shares for any taxable year significantly in excess of any cash distributions received in such taxable year. U.S. Holders should consult their tax advisors regarding making QEF Elections in their particular circumstances.

If we are considered a PFIC, a U.S. Holder will also be subject to annual information reporting requirements. U.S. Holders should consult their tax advisors about the potential application of the PFIC rules to an investment in Class A common shares.

YOU ARE STRONGLY URGED TO CONSULT YOUR TAX ADVISOR REGARDING THE APPLICATION OF THE PFIC RULES TO YOUR INVESTMENT IN OUR Class A common shares.

Information Reporting and Backup Withholding

Dividend payments with respect to Class A common shares and proceeds from the sale, exchange or other disposition of Class A common shares may be subject to information reporting to the IRS and U.S. backup withholding. Certain U.S. Holders are exempt from backup withholding, including corporations and certain tax-exempt organizations. A U.S. Holder will be subject to backup withholding if such holder is not otherwise exempt and such holder:

- fails to furnish the holder's taxpayer identification number, which for an individual is ordinarily his or her social security number;
- furnishes an incorrect taxpayer identification number;
- is notified by the IRS that the holder previously failed to properly report payments of interest or dividends; or
- fails to certify under penalties of perjury that the holder has furnished a correct taxpayer identification number and that the IRS has not notified the holder that the holder is subject to backup withholding.

Backup withholding is not an additional tax. Any amounts withheld under the backup withholding rules may be allowed as a refund or a credit against the U.S. Holder's U.S. federal income tax liability, provided the required information is timely furnished to the IRS. U.S. Holders should consult their tax advisors regarding their qualification for an exemption from backup withholding and the procedures for obtaining such an exemption.

Additional Reporting Requirements

Certain U.S. Holders who are individuals (and certain entities) that hold an interest in "specified foreign financial assets" (which may include the Class A common shares) are required to report information relating to such assets, subject to certain exceptions (including an exception for Class A common shares held in accounts maintained by certain financial institutions). Penalties can apply if U.S. Holders fail to satisfy such reporting requirements. U.S. Holders should consult their tax advisors regarding the applicability of these requirements to their acquisition and ownership of Class A common shares.

UNDERWRITING

The company and the underwriters named below have entered into an underwriting agreement with respect to the shares being offered. Subject to certain conditions, each underwriter has severally agreed to purchase the number of shares indicated in the following table. Goldman Sachs & Co. LLC and J.P. Morgan Securities LLC are the representatives of the underwriters.

Underwriters	Number of Shares
Goldman Sachs & Co. LLC	
J.P. Morgan Securities LLC	
JMP Securities LLC	
Wedbush Securities Inc.	
Total	7,000,000

The underwriters will be committed to take and pay for all of the shares being offered, if any are taken, other than the shares covered by the option described below unless and until this option is exercised.

The underwriters will have an option to buy up to an additional 1,050,000 Class A common shares from the company to cover sales by the underwriters of a greater number of shares than the total number set forth in the table above. They may exercise that option for 30 days. If any shares are purchased pursuant to this option, the underwriters will severally purchase shares in approximately the same proportion as set forth in the table above.

The following table shows the per share and total underwriting discounts and commissions to be paid to the underwriters by the company. Such amounts are shown assuming both no exercise and full exercise of the underwriters' option to purchase 1,050,000 additional Class A common shares.

Paid by the Company

	No Exercise	Full Exercise
Per Share	$	$
Total	$	$

Shares sold by the underwriters to the public will initially be offered at the initial public offering price set forth on the cover of this prospectus. Any shares sold by the underwriters to securities dealers may be sold at a discount of up to $ per share from the initial public offering price. After the initial offering of the shares, the representatives may change the offering price and the other selling terms. The offering of the shares by the underwriters is subject to receipt and acceptance and subject to the underwriters' right to reject any order in whole or in part.

The company and its officers, directors and holders of substantially all of the company's common shares have agreed or will agree with the underwriters, subject to certain exceptions, not to dispose of or hedge any of their common shares or securities convertible into or exchangeable for common shares during the period from the date of this prospectus continuing through the date 180 days after the date of this prospectus, except with the prior written consent of the representatives. This agreement does not apply to any existing employee benefit plans. See the section entitled "Shares Eligible for Future Sale" for a discussion of certain transfer restrictions.

Prior to the offering, there has been no public market for the shares. The initial public offering price has been negotiated among the company and the representatives. Among the factors to be considered in determining the initial public offering price of the shares, in addition to prevailing market conditions, will be the company's historical performance, estimates of the business potential and earnings prospects of the company, an assessment of the company's management and the

consideration of the above factors in relation to market valuation of companies in related businesses.

We have applied to have our Class A common shares listed on The Nasdaq Global Market under the symbol "KNSA."

In connection with the offering, the underwriters may purchase and sell common shares in the open market. These transactions may include short sales, stabilizing transactions and purchases to cover positions created by short sales. Short sales involve the sale by the underwriters of a greater number of shares than they are required to purchase in the offering, and a short position represents the amount of such sales that have not been covered by subsequent purchases. A "covered short position" is a short position that is not greater than the amount of additional Class A common shares for which the underwriters' option described above may be exercised. The underwriters may cover any covered short position by either exercising their option to purchase additional Class A common shares or purchasing shares in the open market. In determining the source of shares to cover the covered short position, the underwriters will consider, among other things, the price of shares available for purchase in the open market as compared to the price at which they may purchase additional Class A common shares pursuant to the option described above. "Naked" short sales are any short sales that create a short position greater than the amount of additional Class A common shares for which the option described above may be exercised. The underwriters must cover any such naked short position by purchasing shares in the open market. A naked short position is more likely to be created if the underwriters are concerned that there may be downward pressure on the price of the common shares in the open market after pricing that could adversely affect investors who purchase in the offering. Stabilizing transactions consist of various bids for or purchases of common shares made by the underwriters in the open market prior to the completion of the offering.

The underwriters may also impose a penalty bid. This occurs when a particular underwriter repays to the underwriters a portion of the underwriting discount received by it because the representatives have repurchased shares sold by or for the account of such underwriter in stabilizing or short covering transactions.

Purchases to cover a short position and stabilizing transactions, as well as other purchases by the underwriters for their own accounts, may have the effect of preventing or retarding a decline in the market price of the company's common shares, and together with the imposition of the penalty bid, may stabilize, maintain or otherwise affect the market price of the common shares. As a result, the price of the common shares may be higher than the price that otherwise might exist in the open market. The underwriters are not required to engage in these activities and may end any of these activities at any time. These transactions may be effected on Nasdaq, in the over-the-counter market or otherwise.

The company estimates that its share of the total expenses of the offering, excluding underwriting discounts and commissions, will be approximately $3.8 million. We will agree to reimburse the underwriters for expenses related to any applicable state securities filings and to the Financial Industry Regulatory Authority incurred by them in connection with this offering in an amount up to $30,000.

The company has agreed to indemnify the several underwriters against certain liabilities, including liabilities under the Securities Act of 1933.

The underwriters and their respective affiliates are full service financial institutions engaged in various activities, which may include sales and trading, commercial and investment banking, advisory, investment management, investment research, principal investment, hedging, market making, brokerage and other financial and non-financial activities and services. Certain of the underwriters and their respective affiliates have provided, and may in the future provide, a variety of

these services to the issuer and to persons and entities with relationships with the issuer, for which they received or will receive customary fees and expenses.

In the ordinary course of their various business activities, the underwriters and their respective affiliates, officers, directors and employees may purchase, sell or hold a broad array of investments and actively trade securities, derivatives, loans, commodities, currencies, credit default swaps and other financial instruments for their own account and for the accounts of their customers, and such investment and trading activities may involve or relate to assets, securities and/or instruments of the issuer (directly, as collateral securing other obligations or otherwise) and/or persons and entities with relationships with the issuer. The underwriters and their respective affiliates may also communicate independent investment recommendations, market color or trading ideas and/or publish or express independent research views in respect of such assets, securities or instruments and may at any time hold, or recommend to clients that they should acquire, long and/or short positions in such assets, securities and instruments.

A prospectus in electronic format may be made available on the web sites maintained by one or more underwriters, or selling group members, if any, participating in the offering. The underwriters may agree to allocate a number of shares to underwriters and selling group members for sale to their online brokerage account holders. Internet distributions will be allocated by the representatives to underwriters and selling group members that may make Internet distributions on the same basis as other allocations.

Sales of shares made outside of the United States may be made by affiliates of the underwriters. Other than in the United States, no action has been taken by us or the underwriters that would permit a public offering of the securities offered by this prospectus in any jurisdiction where action for that purpose is required. The securities offered by this prospectus may not be offered or sold, directly or indirectly, nor may this prospectus or any other offering material or advertisements in connection with the offer and sale of any such securities be distributed or published in any jurisdiction, except under circumstances that will result in compliance with the applicable rules and regulations of that jurisdiction. Persons into whose possession this prospectus comes are advised to inform themselves about and to observe any restrictions relating to the offering and the distribution of this prospectus. This prospectus does not constitute an offer to sell or a solicitation of an offer to buy any securities offered by this prospectus in any jurisdiction in which such an offer or a solicitation is unlawful.

Selling Restrictions

European Economic Area

In relation to each Member State of the European Economic Area which has implemented the Prospectus Directive (each, a "Relevant Member State") an offer to the public of our common shares may not be made in that Relevant Member State, except that an offer to the public in that Relevant Member State of our common shares may be made at any time under the following exemptions under the Prospectus Directive:

- To any legal entity which is a qualified investor as defined in the Prospectus Directive;
- To fewer than 150 natural or legal persons (other than qualified investors as defined in the Prospectus Directive), subject to obtaining the prior consent of the Goldman, Sachs & Co. LLC and J.P. Morgan Securities LLC for any such offer; or
- In any other circumstances falling within Article 3(2) of the Prospectus Directive;

provided that no such offer of our common shares shall result in a requirement for the publication by us or any Brazilian placement agent of a prospectus pursuant to Article 3 of the Prospectus Directive.

For the purposes of this provision, the expression an "offer to public" in relation to our common shares in any Relevant Member State means the communication in any form and by any means of sufficient information on the terms of the offer and our common shares to be offered so as to enable an investor to decide to purchase our common shares, as the same may be varied in that Member State by any measure implementing the Prospectus Directive in that Member State, the expression "Prospectus Directive" means Directive 2003/71/EC (as amended), including by Directive 2010/73/EU and includes any relevant implementing measure in the Relevant Member State.

This European Economic Area selling restriction is in addition to any other selling restrictions set out below.

United Kingdom

In the United Kingdom, this prospectus is only addressed to and directed as qualified investors who are (i) investment professionals falling within Article 19(5) of the Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 (the Order); or (ii) high net worth entities and other persons to whom it may lawfully be communicated, falling within Article 49(2)(a) to (d) of the Order (all such persons together being referred to as "relevant persons"). Any investment or investment activity to which this prospectus relates is available only to relevant persons and will only be engaged with relevant persons. Any person who is not a relevant person should not act or relay on this prospectus or any of its contents.

Canada

The securities may be sold in Canada only to purchasers purchasing, or deemed to be purchasing, as principal that are accredited investors, as defined in National Instrument 45-106 Prospectus Exemptions or subsection 73.3(1) of the Securities Act (Ontario), and are permitted clients, as defined in National Instrument 31-103 Registration Requirements, Exemptions, and Ongoing Registrant Obligations. Any resale of the securities must be made in accordance with an exemption from, or in a transaction not subject to, the prospectus requirements of applicable securities laws.

Securities legislation in certain provinces or territories of Canada may provide a purchaser with remedies for rescission or damages if this offering memorandum (including any amendment thereto) contains a misrepresentation, provided that the remedies for rescission or damages are exercised by the purchaser within the time limit prescribed by the securities legislation of the purchaser's province or territory. The purchaser should refer to any applicable provisions of the securities legislation of the purchaser's province or territory of these rights or consult with a legal advisor.

Pursuant to section 3A.3 of National Instrument 33-105 Underwriting Conflicts (NI 33-105), the underwriters are not required to comply with the disclosure requirements of NI 33-105 regarding underwriter conflicts of interest in connection with this offering.

Hong Kong

The shares may not be offered or sold in Hong Kong by means of any document other than (i) in circumstances which do not constitute an offer to the public within the meaning of the Companies (Winding Up and Miscellaneous Provisions) Ordinance (Cap. 32 of the Laws of Hong Kong) ("Companies (Winding Up and Miscellaneous Provisions) Ordinance") or which do not constitute an invitation to the public within the meaning of the Securities and Futures Ordinance (Cap. 571 of the Laws of Hong Kong) ("Securities and Futures Ordinance"), or (ii) to "professional investors" as defined in the Securities and Futures Ordinance and any rules made thereunder, or (iii) in other circumstances which do not result in the document being a "prospectus" as defined in

the Companies (Winding Up and Miscellaneous Provisions) Ordinance, and no advertisement, invitation or document relating to the shares may be issued or may be in the possession of any person for the purpose of issue (in each case whether in Hong Kong or elsewhere), which is directed at, or the contents of which are likely to be accessed or read by, the public in Hong Kong (except if permitted to do so under the securities laws of Hong Kong) other than with respect to shares which are or are intended to be disposed of only to persons outside Hong Kong or only to "professional investors" in Hong Kong as defined in the Securities and Futures Ordinance and any rules made thereunder.

Singapore

This prospectus has not been registered as a prospectus with the Monetary Authority of Singapore. Accordingly, this prospectus and any other document or material in connection with the offer or sale, or invitation for subscription or purchase, of the shares may not be circulated or distributed, nor may the shares be offered or sold, or be made the subject of an invitation for subscription or purchase, whether directly or indirectly, to persons in Singapore other than (i) to an institutional investor (as defined under Section 4A of the Securities and Futures Act, Chapter 289 of Singapore (the "SFA")) under Section 274 of the SFA, (ii) to a relevant person (as defined in Section 275(2) of the SFA) pursuant to Section 275(1) of the SFA, or any person pursuant to Section 275(1A) of the SFA, and in accordance with the conditions specified in Section 275 of the SFA or (iii) otherwise pursuant to, and in accordance with the conditions of, any other applicable provision of the SFA, in each case subject to conditions set forth in the SFA.

Where the shares are subscribed or purchased under Section 275 of the SFA by a relevant person which is a corporation (which is not an accredited investor (as defined in Section 4A of the SFA)) the sole business of which is to hold investments and the entire share capital of which is owned by one or more individuals, each of whom is an accredited investor, the securities (as defined in Section 239(1) of the SFA) of that corporation shall not be transferable for 6 months after that corporation has acquired the shares under Section 275 of the SFA except: (1) to an institutional investor under Section 274 of the SFA or to a relevant person (as defined in Section 275(2) of the SFA), (2) where such transfer arises from an offer in that corporation's securities pursuant to Section 275(1A) of the SFA, (3) where no consideration is or will be given for the transfer, (4) where the transfer is by operation of law, (5) as specified in Section 276(7) of the SFA, or (6) as specified in Regulation 32 of the Securities and Futures (Offers of Investments) (Shares and Debentures) Regulations 2005 of Singapore ("Regulation 32").

Where the shares are subscribed or purchased under Section 275 of the SFA by a relevant person which is a trust (where the trustee is not an accredited investor (as defined in Section 4A of the SFA)) whose sole purpose is to hold investments and each beneficiary of the trust is an accredited investor, the beneficiaries' rights and interest (howsoever described) in that trust shall not be transferable for 6 months after that trust has acquired the shares under Section 275 of the SFA except: (1) to an institutional investor under Section 274 of the SFA or to a relevant person (as defined in Section 275(2) of the SFA), (2) where such transfer arises from an offer that is made on terms that such rights or interest are acquired at a consideration of not less than $200,000 (or its equivalent in a foreign currency) for each transaction (whether such amount is to be paid for in cash or by exchange of securities or other assets), (3) where no consideration is or will be given for the transfer, (4) where the transfer is by operation of law, (5) as specified in Section 276(7) of the SFA, or (6) as specified in Regulation 32.

Japan

The securities have not been and will not be registered under the Financial Instruments and Exchange Act of Japan (Act No. 25 of 1948, as amended), or the FIEA. The securities may not be offered or sold, directly or indirectly, in Japan or to or for the benefit of any resident of Japan (including any person resident in Japan or any corporation or other entity organized under the laws of Japan) or to others for reoffering or resale, directly or indirectly, in Japan or to or for the benefit of any resident of Japan, except pursuant to an exemption from the registration requirements of the FIEA and otherwise in compliance with any relevant laws and regulations of Japan.

LEGAL MATTERS

The validity of the Class A common shares offered hereby will be passed upon for us by Conyers Dill & Pearman Limited. Certain legal matters as to U.S. law in connection with this offering will be passed upon for us by Latham & Watkins LLP. Certain legal matters will be passed upon for the underwriters by Ropes & Gray LLP.

EXPERTS

The financial statements as of December 31, 2016 and 2017 and for the years then ended included in this prospectus have been so included in reliance on the report of PricewaterhouseCoopers LLP, an independent registered public accounting firm, given on the authority of said firm as experts in auditing and accounting.

EXCHANGE CONTROLS

The permission of the Bermuda Monetary Authority is required, pursuant to the provisions of the Exchange Control Act 1972 and related regulations, for all issuances and transfers of shares (which includes our Class A common shares) of Bermuda companies to or from a non-resident of Bermuda for exchange control purposes, other than in cases where the Bermuda Monetary Authority has granted a general permission. The Bermuda Monetary Authority has, pursuant to its statement of June 1, 2005, given its general permission under the Exchange Control Act 1972 (and related regulations) for the issue and free transferability of our Class A common shares to and between non-residents of Bermuda for exchange control purposes, provided that the Class A common shares remain listed on an appointed stock exchange, which includes The Nasdaq Global Market. Approvals or permissions given by the Bermuda Monetary Authority do not constitute a guarantee by the Bermuda Monetary Authority as to our performance or our creditworthiness. Accordingly, in giving such consent or permissions, the Bermuda Monetary Authority shall not be liable for the financial soundness, performance or default of our business or for the correctness of any opinions or statements expressed herein. Certain issues and transfers of Class A common shares involving persons deemed resident in Bermuda for exchange control purposes require the specific consent of the Bermuda Monetary Authority.

ENFORCEMENT OF CIVIL LIABILITIES UNDER UNITED STATES FEDERAL SECURITIES LAWS

We are organized pursuant to the laws of Bermuda. In addition, it is anticipated that some or all of our directors and officers will reside outside the United States, and all or a substantial portion of our assets and their assets are or may be located in jurisdictions outside the United States. As a result, it may be difficult for you to effect service of process within the United States upon those persons or us or to recover against them or us on judgments of United States courts, including judgments predicated upon civil liability provisions of the United States federal securities laws.

We have been advised that there is no treaty in force between the United States and Bermuda providing for the reciprocal recognition and enforcement of judgments in civil and commercial matters. As a result, whether a U.S. judgment would be enforceable in Bermuda against us or our directors and officers depends on whether the U.S. court that entered the judgment is recognized by the Bermuda court as having jurisdiction over us or our directors and officers, as determined by reference to Bermuda conflict of law rules. A judgment debt from a U.S. court that is final and for a sum certain based on U.S. federal securities laws will not be enforceable in Bermuda unless the judgment debtor had submitted to the jurisdiction of the U.S. court, and the issue of submission and jurisdiction is a matter of Bermuda (not U.S.) law.

In addition, and irrespective of jurisdictional issues, the Bermuda courts will not enforce a U.S. federal securities law that is either penal or contrary to Bermuda public policy. We have been advised that an action brought pursuant to a public or penal law, the purpose of which is the

enforcement of a sanction, power or right at the instance of the state in its sovereign capacity, will not be entertained by a Bermuda court. Certain remedies available under the laws of U.S. jurisdictions, including certain remedies under U.S. federal securities laws, would not be available under Bermuda law or enforceable in a Bermuda court, as they would be contrary to Bermuda public policy. Further, no claim may be brought in Bermuda against us or our directors and officers in the first instance for violation of U.S. federal securities laws because these laws have no extraterritorial jurisdiction under Bermuda law and do not have force of law in Bermuda. A Bermuda court may, however, impose civil liability on us or our directors and officers if the facts alleged in a complaint constitute or give rise to a cause of action under Bermuda law.

WHERE YOU CAN FIND MORE INFORMATION

We have filed with the SEC a registration statement on Form S-1 under the Securities Act with respect to the Class A common shares offered hereby. This prospectus, which constitutes a part of the registration statement, does not contain all of the information set forth in the registration statement or the exhibits and schedules filed therewith. For further information about us and the Class A common shares offered hereby, we refer you to the registration statement and the exhibits and schedules filed thereto. Statements contained in this prospectus regarding the contents of any contract or any other document that is filed as an exhibit to the registration statement are not necessarily complete, and each such statement is qualified in all respects by reference to the full text of such contract or other document filed as an exhibit to the registration statement. Upon the closing of this offering, we will be required to file periodic reports, proxy statements, and other information with the SEC pursuant to the Securities Exchange Act of 1934. You may read and copy this information at the Public Reference Room of the SEC, 100 F Street, N.E., Room 1580, Washington, D.C. 20549. You may obtain information on the operation of the public reference rooms by calling the SEC at 1-800-SEC-0330. The SEC also maintains an Internet website that contains reports, proxy statements and other information about registrants, like us, that file electronically with SEC. The address of that site is *www.sec.gov.*

Upon the closing of this offering, we will become subject to the information and periodic reporting requirements of the Exchange Act and, in accordance therewith, we will file periodic reports, proxy statements and other information with the SEC. Such periodic reports, proxy statements and other information will be available for inspection and copying at the public reference room and website of the SEC referred to above.

INDEX TO CONSOLIDATED FINANCIAL STATEMENTS

REPORT OF INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM

To the Board of Directors and Shareholders of
Kiniksa Pharmaceuticals, Ltd.

Opinion on the Financial Statements

We have audited the accompanying consolidated balance sheets of Kiniksa Pharmaceuticals, Ltd. and its subsidiary (the "Company") as of December 31, 2017 and 2016, and the related consolidated statements of operations and comprehensive loss, of convertible preferred shares and shareholders' deficit and of cash flows for the years then ended, including the related notes (collectively referred to as the "consolidated financial statements"). In our opinion, the consolidated financial statements present fairly, in all material respects, the financial position of the Company as of December 31, 2017 and 2016, and the results of its operations and its cash flows for the years then ended in conformity with accounting principles generally accepted in the United States of America.

Basis for Opinion

These consolidated financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on the Company's consolidated financial statements based on our audits. We are a public accounting firm registered with the Public Company Accounting Oversight Board (United States) ("PCAOB") and are required to be independent with respect to the Company in accordance with the U.S. federal securities laws and the applicable rules and regulations of the Securities and Exchange Commission and the PCAOB.

We conducted our audits of these consolidated financial statements in accordance with the standards of the PCAOB. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the consolidated financial statements are free of material misstatement, whether due to error or fraud.

Our audits included performing procedures to assess the risks of material misstatement of the consolidated financial statements, whether due to error or fraud, and performing procedures that respond to those risks. Such procedures included examining, on a test basis, evidence regarding the amounts and disclosures in the consolidated financial statements. Our audits also included evaluating the accounting principles used and significant estimates made by management, as well as evaluating the overall presentation of the consolidated financial statements. We believe that our audits provide a reasonable basis for our opinion.

/s/ PricewaterhouseCoopers LLP

Boston, Massachusetts
February 27, 2018, except for the effects of the reverse share split discussed in Note 14 to the consolidated financial statements, as to which the date is May 14, 2018

We have served as the Company's auditor since 2016.

KINIKSA PHARMACEUTICALS, LTD.

CONSOLIDATED BALANCE SHEETS

(In thousands, except share and per share amounts)

	December 31, 2016	December 31, 2017	March 31, 2018	Pro Forma March 31, 2018
			(unaudited)	
Assets				
Current assets:				
Cash and cash equivalents	$ 55,970	$ 45,555	$ 221,108	$ 221,108
Restricted cash	105	105	105	105
Prepaid expenses and other current assets	259	1,444	1,240	1,240
Total current assets	56,334	47,104	222,453	222,453
Property and equipment, net	84	125	126	126
Restricted cash	—	—	210	210
Deferred offering costs	8	25	1,608	1,608
Deferred tax assets	41	238	378	378
Total assets	$ 56,467	$ 47,492	$ 224,775	$ 224,775
Liabilities, Convertible Preferred Shares and Shareholders' Equity (Deficit)				
Current liabilities:				
Accounts payable	$ 212	$ 1,218	$ 2,796	$ 2,796
Accrued expenses	2,090	6,212	6,472	6,472
Accrued milestone	—	10,000	10,000	10,000
Total current liabilities	2,302	17,430	19,268	19,268
Deferred rent	—	—	30	30
Total liabilities	2,302	17,430	19,298	19,298
Commitments and contingencies (Note 12)				
Convertible preferred shares (Series A, B and C), $0.000273235 par value; 17,128,120 shares, 22,885,492 shares and 35,670,093 shares designated, issued and outstanding as of December 31, 2016 and 2017 and March 31, 2018 (unaudited), respectively; aggregate liquidation preference of $80,000, $120,000 and $320,000 as of December 31, 2016 and 2017 and March 31, 2018 (unaudited), no shares issued or outstanding, pro forma as of March 31, 2018 (unaudited)	79,897	119,770	310,592	—
Shareholders' equity (deficit):				
Class A common shares, par value of $0.000273235 per share; 4,491,921 shares designated as of December 31, 2016 and 5,507,938 shares designated as of December 31, 2017 and March 31, 2018 (unaudited); 719,976 shares issued and outstanding as of December 31, 2016 and 2017 and 724,550 shares issued and outstanding as of March 31, 2018 (unaudited), 6,270,569 shares issued and outstanding, pro forma, as of March 31, 2018 (unaudited)	—	—	—	2
Class B common shares, par value of $0.000273235 per share; 3,568,353 shares designated, issued and outstanding as of December 31, 2016 and 2017 and March 31, 2018 (unaudited), 4,638,855 shares issued and outstanding, pro forma, as of March 31, 2018 (unaudited)	1	1	1	1
Class A1 common shares, $0.000273235 par value; no shares designated, issued or outstanding as of December 31, 2016 and 2017 and March 31, 2018 (unaudited); 12,995,954 shares issued and outstanding, pro forma as of March 31, 2018 (unaudited)	—	—	—	4
Class B1 common shares, $0.000273235 par value; no shares designated, issued or outstanding as of December 31, 2016 and 2017 and March 31, 2018 (unaudited); 16,057,618 shares issued and outstanding, pro forma as of March 31, 2018 (unaudited)	—	—	—	4
Additional paid-in capital	392	1,289	1,864	312,446
Accumulated deficit	(26,125)	(90,998)	(106,980)	(106,980)
Total shareholders' equity (deficit)	(25,732)	(89,708)	(105,115)	205,477
Total liabilities, convertible preferred shares and shareholders' equity (deficit)	$ 56,467	$ 47,492	$ 224,775	$ 224,775

The accompanying notes are an integral part of these consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.

CONSOLIDATED STATEMENTS OF OPERATIONS AND COMPREHENSIVE LOSS

(In thousands, except share and per share amounts)

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
			(unaudited)	
Operating expenses:				
Research and development	$ 17,439	$ 56,357	$ 3,145	$ 12,630
General and administrative	6,563	9,043	1,903	3,710
Total operating expenses	24,002	65,400	5,048	16,340
Loss from operations	(24,002)	(65,400)	(5,048)	(16,340)
Interest income	65	529	74	305
Loss before benefit (provision) for income taxes	(23,937)	(64,871)	(4,974)	(16,035)
Benefit (provision) for income taxes	(36)	(2)	33	53
Net loss and comprehensive loss	$ (23,973)	$ (64,873)	$ (4,941)	$ (15,982)
Net loss per share attributable to common shareholders—basic and diluted	$ (91.61)	$ (35.85)	$ (3.52)	$ (6.45)
Weighted average common shares outstanding—basic and diluted	261,695	1,809,751	1,405,400	2,478,903
Pro forma net loss per share attributable to common shareholders—basic and diluted (unaudited)		$ (2.74)		$ (0.49)
Pro forma weighted average common shares outstanding—basic and diluted (unaudited)		23,638,410		32,466,951

The accompanying notes are an integral part of these consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.

CONSOLIDATED STATEMENTS OF CONVERTIBLE PREFERRED SHARES AND SHAREHOLDERS' DEFICIT

(In thousands, except share amounts)

	Convertible Preferred Shares (Series A, B and C)		Common Shares (Class A and B)		Additional Paid-In	Accumulated	Total Shareholders'
	Shares	Amount	Shares	Amount	Capital	Deficit	Deficit
Balances at December 31, 2015	8,028,809	$ 37,398	4,282,020	$ 1	$ 14	$ (2,152)	$ (2,137)
Issuance of Series A convertible preferred shares, net of issuance costs of $1	9,099,311	42,499	—	—	—	—	—
Exercise of options	—	—	6,309	—	10	—	10
Share-based compensation expense	—	—	—	—	368	—	368
Net loss	—	—	—	—	—	(23,973)	(23,973)
Balances at December 31, 2016	17,128,120	79,897	4,288,329	1	392	(26,125)	(25,732)
Issuance of Series B convertible preferred shares, net of issuance costs of $127	5,757,372	39,873	—	—	—	—	—
Share-based compensation expense	—	—	—	—	897	—	897
Net loss	—	—	—	—	—	(64,873)	(64,873)
Balances at December 31, 2017	22,885,492	119,770	4,288,329	1	1,289	(90,998)	(89,708)
Issuance of Series C convertible preferred shares, net of issuance costs of $9,178	12,784,601	190,822	—	—	—	—	—
Exercise of options	—	—	4,574	—	17	—	17
Share-based compensation expense	—	—	—	—	558	—	558
Net loss	—	—	—	—	—	(15,982)	(15,982)
Balances at March 31, 2018 (unaudited)	35,670,093	$ 310,592	4,292,903	$ 1	$ 1,864	$ (106,980)	$ (105,115)

The accompanying notes are an integral part of these consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.

CONSOLIDATED STATEMENTS OF CASH FLOWS

(In thousands)

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
				(unaudited)
Cash flows from operating activities:				
Net loss	$ (23,973)	$ (64,873)	$ (4,941)	$ (15,982)
Adjustments to reconcile net loss to net cash used in operating activities:				
Depreciation expense	22	28	6	8
Share-based compensation expense	368	897	114	558
Loss on disposal of property and equipment	—	—	—	66
Non-cash rent expense	—	—	—	30
Deferred income taxes	(46)	(197)	(34)	(140)
Changes in operating assets and liabilities:				
Prepaid expenses and other current assets	(219)	(1,185)	(122)	204
Accounts payable	119	1,006	375	1,091
Accrued expenses	1,862	4,105	(1,078)	(807)
Accrued milestone	—	10,000	—	—
Net cash used in operating activities	(21,867)	(50,219)	(5,679)	(14,972)
Cash flows from investing activities:				
Purchases of property and equipment	(3)	(69)	(18)	(75)
Net cash used in investing activities	(3)	(69)	(18)	(75)
Cash flows from financing activities:				
Proceeds from issuance of Series A convertible preferred shares, net of issuance costs	42,499	—	—	—
Proceeds from issuance of Series B convertible preferred shares, net of issuance costs	—	39,873	39,873	—
Proceeds from issuance of Series C convertible preferred shares, net of issuance costs	—	—	—	190,822
Payments of deferred offering costs	—	—	—	(29)
Proceeds from exercise of options	10	—	—	17
Net cash provided by financing activities	42,509	39,873	39,873	190,810
Net increase (decrease) in cash and cash equivalents and restricted cash	20,639	(10,415)	34,176	175,763
Cash and cash equivalents and restricted cash at beginning of period	35,436	56,075	56,075	45,660
Cash and cash equivalents and restricted cash at end of period	$ 56,075	$ 45,660	$ 90,251	$ 221,423
Supplemental information:				
Cash paid for income taxes	$ 115	$ 290	$ —	$ —
Supplemental disclosure of non-cash investing and financing activities:				
Deferred offering costs included in accrued expenses and accounts payable	$ 8	$ 25	$ —	$ 1,579

The accompanying notes are an integral part of these consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS

(Amounts in thousands, except share and per share amounts)

1. Nature of the Business and Basis of Presentation

Kiniksa Pharmaceuticals, Ltd. (the "Company") is a clinical-stage biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. The Company was incorporated in July 2015 as a Bermuda exempted company. The Company has a pipeline of product candidates across various stages of development, currently focused on autoinflammatory and autoimmune conditions.

The Company is subject to risks and uncertainties common to early-stage companies in the biotechnology industry. There can be no assurance that the Company's research and development will be successfully completed, that adequate protection for the Company's technology will be obtained, that any products developed will obtain necessary government regulatory approval or that any products, if approved, will be commercially viable. The Company operates in an environment of rapid technological innovation and substantial competition from pharmaceutical and biotechnological companies. In addition, the Company is dependent upon the services of its employees, consultants and service providers. Even if the Company's product development efforts are successful, it is uncertain when, if ever, the Company will realize significant revenue from product sales.

Through December 31, 2017 and March 31, 2018 (unaudited), the Company has funded its operations primarily with proceeds from the sale of convertible preferred shares. The Company has incurred recurring losses since its inception, including net losses of $23,973 and $64,873 for the years ended December 31, 2016 and 2017, respectively and $4,941 and $15,982 for the three months ended March 31, 2017 and 2018 (unaudited), respectively. In addition, as of December 31, 2017 and March 31, 2018 (unaudited), the Company had an accumulated deficit of $90,998 and $106,980, respectively. The Company expects to continue to generate operating losses for the foreseeable future. As of February 27, 2018, the issuance date of the annual consolidated financial statements for the years ended December 31, 2016 and 2017, the Company expected that its cash and cash equivalents of $45,555 as of December 31, 2017, together with the $200,000 of gross proceeds received from the Company's sale of Series C convertible preferred shares in February 2018 (see Note 6), would be sufficient to fund its operating expenses and capital expenditure requirements for at least twelve months from the issuance date of the annual consolidated financial statements. As of April 27, 2018, the issuance date of the interim consolidated financial statements for the three months ended March 31, 2018, the Company expected that its cash and cash equivalents of $221,108 as of March 31, 2018 (unaudited) would be sufficient to fund its operating expenses and capital expenditure requirements for at least twelve months from the issuance date of the interim consolidated financial statements. The future viability of the Company beyond that point is dependent on its ability to raise additional capital to finance its operations.

The Company is seeking to complete an initial public offering ("IPO") of its common shares. Upon the closing of a qualified public offering, on specified terms, the Company's outstanding convertible preferred shares will automatically convert into common shares (see Note 6). In the event the Company does not complete an IPO, the Company expects to seek additional funding through private equity financings, debt financings, or other capital sources, which may include collaborations with other companies, government funding arrangements or other strategic transactions. The Company may not be able to obtain financing on acceptable terms, or at all. The

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

1. Nature of the Business and Basis of Presentation (Continued)

terms of any financing may adversely affect the holdings or the rights of the Company's shareholders.

If the Company is unable to obtain funding, the Company will be forced to delay, reduce or eliminate some or all of its research and development programs, product portfolio expansion or commercialization efforts, which could adversely affect its business prospects, or the Company may be unable to continue operations. Although management continues to pursue these plans, there is no assurance that the Company will be successful in obtaining sufficient funding on terms acceptable to the Company to fund continuing operations, if at all.

2. Summary of Significant Accounting Policies

Principles of Consolidation

The accompanying consolidated financial statements have been prepared in accordance with accounting principles generally accepted in the United States of America ("GAAP") and include the accounts of the Company and its wholly owned U.S. subsidiary, Kiniksa Pharmaceuticals Corp. ("Kiniksa US"), after elimination of all significant intercompany accounts and transactions.

In assessing the consolidation requirement for variable interest entities ("VIEs"), the Company focuses on identifying whether it has both the power to direct the activities that most significantly impact the VIE's economic performance and the obligation to absorb losses or the right to receive benefits from the VIE. In the event that the Company is the primary beneficiary of a VIE, the assets, liabilities, and results of operations of the VIE would be included in the Company's consolidated financial statements. At December 31, 2016 and 2017 and during the years then ended and at March 31, 2018 and during the three months then ended (unaudited), the Company was not the primary beneficiary of a VIE.

Use of Estimates

The preparation of the Company's consolidated financial statements in conformity with GAAP requires management to make estimates and assumptions that affect the reported amounts of assets and liabilities, the disclosure of contingent assets and liabilities at the date of the consolidated financial statements and the reported amounts of expenses during the reporting period. Significant estimates and assumptions reflected in these consolidated financial statements include, but are not limited to, the accrual for research and development expenses and the valuation of common shares and share-based awards. Estimates are periodically reviewed in light of changes in circumstances, facts and experience. Changes in estimates are recorded in the period in which they become known. Actual results could differ from those estimates.

Unaudited Interim Financial Information

The accompanying consolidated balance sheet as of March 31, 2018, the consolidated statements of operations and comprehensive loss and of cash flows for the three months ended March 31, 2017 and 2018, and the consolidated statement of convertible preferred shares and shareholders' deficit for the three months ended March 31, 2018 are unaudited. The unaudited interim consolidated financial statements have been prepared on the same basis as the audited

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

annual consolidated financial statements and, in the opinion of management, reflect all adjustments, which include only normal recurring adjustments, necessary for the fair statement of the Company's financial position as of March 31, 2018 and the results of its operations and its cash flows for the three months ended March 31, 2017 and 2018. The financial data and other information disclosed in these notes related to the three months ended March 31, 2017 and 2018 are also unaudited. The results for the three months ended March 31, 2018 are not necessarily indicative of results to be expected for the year ending December 31, 2018, any other interim periods, or any future year or period.

Unaudited Pro Forma Information

The accompanying unaudited pro forma consolidated balance sheet as of March 31, 2018 has been prepared to give effect, upon the closing of a qualified IPO, to the automatic conversion of (i) all outstanding Series A convertible preferred shares into 1,070,502 Class B common shares and 16,057,618 Class B1 common shares and (ii) all outstanding Series B and Series C convertible preferred shares into 5,546,019 Class A common shares and 12,995,954 Class A1 common shares as if the Company's proposed IPO had occurred on March 31, 2018.

In the accompanying consolidated statements of operations and comprehensive loss, the unaudited pro forma basic and diluted net loss per share attributable to common shareholders for the year ended December 31, 2017 and the three months ended March 31, 2018 have been prepared to give effect, upon the closing of a qualified IPO, to the automatic conversion of (i) all outstanding Series A convertible preferred shares into 1,070,502 Class B common shares and 16,057,618 Class B1 common shares and (ii) all outstanding Series B and Series C convertible preferred shares into 5,546,019 Class A common shares and 12,995,954 Class A1 common shares as if the proposed IPO had occurred on the later of January 1, 2017 or the issuance date of the convertible preferred shares.

Cash and Cash Equivalents

The Company classifies deposits in banks, money market funds and cash invested temporarily in various instruments with maturities of three months or less at the time of purchase as cash and cash equivalents. At December 31, 2016 and 2017 and March 31, 2018 (unaudited), cash and cash equivalents consisted principally of U.S. Treasury notes, amounts held in money market accounts and cash on deposit at commercial banks.

Concentrations of Credit Risk

Financial instruments that potentially subject the Company to concentrations of credit risk consist principally of cash and cash equivalents. At December 31, 2016 and 2017 and March 31, 2018 (unaudited), all of the Company's cash and cash equivalents were held at two financial institutions. The Company generally maintains balances in various operating accounts at financial institutions that management believes to be of high credit quality, in amounts that may exceed federally insured limits. The Company has not experienced any losses related to its cash and cash equivalents and does not believe that it is subject to unusual credit risk beyond the normal credit risk associated with commercial banking relationships.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

Restricted Cash

Restricted cash classified as a current asset as of December 31, 2016 and 2017 and March 31, 2018 (unaudited) includes cash held in a money market fund in connection with the Company's corporate credit cards. These amounts have been classified as current assets based on the contractual release date of the restrictions.

In conjunction with the Company's lease agreement entered into in March 2018 (see Note 12), the Company maintains a letter of credit for the benefit of the landlord. As of March 31, 2018 (unaudited), the underlying cash balance of $210 securing this letter of credit, was classified as non-current in the consolidated balance sheet.

Property and Equipment

Property and equipment are recorded at cost and depreciated over the estimated useful lives of the related assets using the straight-line method. Maintenance and repairs are charged to expense as incurred. When assets are retired or otherwise disposed of, the cost of these assets and related accumulated depreciation or amortization are eliminated from the consolidated balance sheet and any resulting gains or losses are included in the consolidated statement of operations and comprehensive loss in the period of disposal. The expected useful lives of the respective assets are as follows:

	Estimated Useful Life
Computer hardware and software	3 - 5 years
Vehicles	5 years
Laboratory and facility equipment	5 years
Furniture and fixtures	5 - 7 years
Leasehold improvements	Shorter of estimated useful life or lease term

Impairment of Long-Lived Assets

Long-lived assets consist of property and equipment. Long-lived assets to be held and used are tested for recoverability whenever events or changes in business circumstances indicate that the carrying amount of the assets may not be fully recoverable. Factors that the Company considers in deciding when to perform an impairment review include significant underperformance of the business in relation to expectations, significant negative industry or economic trends and significant changes or planned changes in the use of the assets. If an impairment review is performed to evaluate a long-lived asset group for recoverability, the Company compares forecasts of undiscounted cash flows expected to result from the use and eventual disposition of the long-lived asset group to its carrying value. An impairment loss would be recognized when estimated undiscounted future cash flows expected to result from the use of an asset group are less than its carrying amount. The impairment loss would be based on the excess of the carrying value of the impaired asset group over its fair value, determined based on discounted cash flows. To date, the Company has not recorded any impairment losses on long-lived assets.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

Deferred Offering Costs

The Company capitalizes certain legal, professional accounting and other third-party fees that are directly associated with in-process preferred share or common equity financings as deferred offering costs until such financings are consummated. After consummation of the equity financing, these costs are recorded as a reduction to the carrying value of convertible preferred shares or in shareholders' equity (deficit) as a reduction of additional paid-in capital generated as a result of the offering. Should an in-process equity financing be abandoned, the deferred offering costs will be expensed immediately as a charge to operating expenses in the consolidated statements of operations and comprehensive loss. The Company recorded deferred offering costs related to the sale of convertible preferred shares of $8 and $25 as of December 31, 2016 or 2017, respectively. The Company recorded deferred offering costs in contemplation of an initial public offering of its Class A common shares of $1,608 as of March 31, 2018 (unaudited).

Fair Value Measurements

Certain assets and liabilities of the Company are carried at fair value under GAAP. Fair value is defined as the exchange price that would be received for an asset or paid to transfer a liability (an exit price) in the principal or most advantageous market for the asset or liability in an orderly transaction between market participants on the measurement date. Valuation techniques used to measure fair value must maximize the use of observable inputs and minimize the use of unobservable inputs. Financial assets and liabilities carried at fair value are to be classified and disclosed in one of the following three levels of the fair value hierarchy, of which the first two are considered observable and the last is considered unobservable:

- Level 1 — Quoted prices in active markets for identical assets or liabilities.
- Level 2 — Observable inputs (other than Level 1 quoted prices), such as quoted prices in active markets for similar assets or liabilities, quoted prices in markets that are not active for identical or similar assets or liabilities, or other inputs that are observable or can be corroborated by observable market data.
- Level 3 — Unobservable inputs that are supported by little or no market activity that are significant to determining the fair value of the assets or liabilities, including pricing models, discounted cash flow methodologies and similar techniques.

The Company's restricted cash, which is held in a money market fund, is carried at fair value, determined based on Level 1 inputs in the fair value hierarchy described above (see Note 3). The Company's cash equivalents, consisting of money market accounts and U.S. Treasury notes, are carried at fair value, determined based on Level 1 and 2 inputs in the fair value hierarchy described above (see Note 3). The carrying values of the Company's prepaid expenses and other current assets, accounts payable and accrued expenses approximate their fair values due to the short-term nature of these assets and liabilities.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

Segment Information

The Company manages its operations as a single operating segment for the purposes of assessing performance and making operating decisions. The Company's singular focus is on developing and delivering therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need.

Research and Development Costs

Research and development costs are expensed as incurred. Research and development expenses consist of costs incurred to discover, research and develop drug candidates, including personnel expenses, share-based compensation expense, allocated facility-related and depreciation expenses, third-party license fees and external costs of outside vendors engaged to conduct clinical development activities and clinical trials as well as to manufacture clinical trial materials. Non-refundable prepayments for goods or services that will be used or rendered for future research and development activities are recorded as prepaid expenses. Such amounts are recognized as an expense as the goods are delivered or the related services are performed, or until it is no longer expected that the goods will be delivered or the services rendered.

Research Contract Costs and Accruals

The Company has entered into various research and development-related contracts with companies both inside and outside of the United States. The related costs are recorded as research and development expenses as incurred. The Company records accruals for estimated ongoing research costs. When evaluating the adequacy of the accrued liabilities, the Company analyzes progress of the studies or clinical trials, including the phase or completion of events, invoices received and contracted costs. Significant judgments and estimates are made in determining the accrued balances at the end of any reporting period. Actual results could differ materially from the Company's estimates. The Company's historical accrual estimates have not been materially different from the actual costs.

Patent Costs

The Company charges patent-related costs in connection with filing and prosecuting patent applications to operations as incurred as their realization is uncertain. These costs are classified as general and administrative expenses.

Share-Based Compensation

The Company measures all share-based awards granted to employees and directors based on their fair value on the date of the grant and recognizes compensation expense for those awards over the requisite service period, which is generally the vesting period of the respective award. Forfeitures are accounted for as they occur. The Company issues share-based awards with only service-based vesting conditions and records the expense for these awards using the straight-line method. The Company has not issued any share-based awards with performance-based vesting conditions.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

For share-based awards granted to consultants and non-employees, compensation expense is recognized over the vesting period of the awards, which is generally the period during which services are rendered by such consultants and non-employees until completed. At the end of each financial reporting period prior to completion of the service, the fair value of these awards is remeasured using the then-current fair value of the Company's Class A common shares and updated assumption inputs in the Black-Scholes option-pricing model.

The Company classifies share-based compensation expense in its consolidated statements of operations and comprehensive loss in the same manner in which the award recipient's payroll costs are classified or in which the award recipient's service payments are classified.

The fair value of each restricted share award is estimated on the date of grant based on the fair value of the Company's Class A or Class B common shares on that same date. The fair value of each option grant is estimated on the date of grant using the Black-Scholes option-pricing model, which requires inputs based on certain subjective assumptions, including the expected share price volatility, the expected term of the award, the risk-free interest rate, and expected dividends (see Note 8). The Company historically has been a private company and lacks company-specific historical and implied volatility information for its shares. Therefore, it estimates its expected share price volatility based on the historical volatility of publicly traded peer companies and expects to continue to do so until such time as it has adequate historical data regarding the volatility of its own traded share price. The expected term of the Company's options has been determined utilizing the "simplified" method for awards that qualify as "plain-vanilla" options. The expected term of options granted to non-employees is equal to the contractual term of the option award. The risk-free interest rate is determined by reference to the U.S. Treasury yield curve in effect at the time of grant of the award for time periods approximately equal to the expected term of the award. Expected dividend yield is based on the fact that the Company has never paid cash dividends on common shares and does not expect to pay any cash dividends in the foreseeable future.

Comprehensive Loss

Comprehensive loss includes net loss as well as other changes in shareholders' equity (deficit) that result from transactions and economic events other than those with shareholders. There was no difference between net loss and comprehensive loss for each of the periods presented in the accompanying consolidated financial statements.

Income Taxes

The Company accounts for income taxes using the asset and liability method, which requires the recognition of deferred tax assets and liabilities for the expected future tax consequences of events that have been recognized in the consolidated financial statements or in the Company's tax returns. Deferred tax assets and liabilities are determined based on the difference between the financial statement and tax basis of assets and liabilities using enacted tax rates in effect for the year in which the differences are expected to reverse. Changes in deferred tax assets and liabilities are recorded in the provision for income taxes. The Company assesses the likelihood that its deferred tax assets will be recovered from future taxable income and, to the extent it believes, based upon the weight of available evidence, that it is more likely than not that all or a portion of

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

the deferred tax assets will not be realized, a valuation allowance is established through a charge to income tax expense. Potential for recovery of deferred tax assets is evaluated by estimating the future taxable profits expected and considering prudent and feasible tax planning strategies.

The Company accounts for uncertainty in income taxes recognized in the consolidated financial statements by applying a two-step process to determine the amount of tax benefit to be recognized. First, the tax position must be evaluated to determine the likelihood that it will be sustained upon external examination by the taxing authorities. If the tax position is deemed more-likely-than-not to be sustained, the tax position is then assessed to determine the amount of benefit to recognize in the consolidated financial statements. The amount of the benefit that may be recognized is the largest amount that has a greater than 50% likelihood of being realized upon ultimate settlement. The provision for income taxes includes the effects of any resulting tax reserves, or unrecognized tax benefits, that are considered appropriate as well as the related net interest and penalties. To date, the Company has not taken any uncertain tax positions or recorded any reserves, interest or penalties.

Net Income (Loss) per Share

The Company follows the two-class method when computing net income (loss) per share as the Company has issued shares that meet the definition of participating securities. The two-class method determines net income (loss) per share for each class of common and participating securities according to dividends declared or accumulated and participation rights in undistributed earnings. The two-class method requires income available to common shareholders for the period to be allocated between common and participating securities based upon their respective rights to receive dividends as if all income for the period had been distributed.

Basic net income (loss) per share attributable to common shareholders is computed by dividing the net income (loss) attributable to common shareholders by the weighted average number of common shares outstanding for the period. Diluted net income (loss) attributable to common shareholders is computed by adjusting net income (loss) attributable to common shareholders to reallocate undistributed earnings based on the potential impact of dilutive securities. Diluted net income (loss) per share attributable to common shareholders is computed by dividing the diluted net income (loss) attributable to common shareholders by the weighted average number of common shares outstanding for the period, including potential dilutive common shares. For purpose of this calculation, outstanding options, unvested restricted common shares and convertible preferred shares are considered potential dilutive common shares.

The Company's convertible preferred shares contractually entitle the holders of such shares to participate in dividends but do not contractually require the holders of such shares to participate in losses of the Company. Accordingly, in periods in which the Company reports a net loss attributable to common shareholders, such losses are not allocated to such participating securities. In periods in which the Company reports a net loss attributable to common shareholders, diluted net loss per share attributable to common shareholders is the same as basic net loss per share attributable to common shareholders, since dilutive common shares are not assumed to have been issued if their effect is anti-dilutive. The Company reported a net loss attributable to common

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

shareholders for the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited).

Recently Adopted Accounting Pronouncements

In May 2017, the Financial Accounting Standards Board ("FASB") issued Accounting Standards Update ("ASU") No. 2017-09, Compensation — Stock Compensation (Topic 718): Scope of Modification Accounting ("ASU 2017-09"), which clarifies when to account for a change to the terms or conditions of a share-based payment award as a modification. Under the new guidance, modification accounting is required only if the fair value, the vesting conditions or the classification of the award (as equity or liability) changes as a result of the change in terms or conditions. ASU 2017-09 is effective for annual periods beginning after December 15, 2017, including interim periods within those fiscal years. Early adoption is permitted. The Company adopted ASU 2017-09 as of the required effective date of January 1, 2018. The adoption of ASU 2017-09 will have an impact on the modification of stock-based awards, if any, after the date of adoption. The adoption of ASU 2017-09 did not have an impact on the Company's financial position, results of operations or cash flows.

In January 2017, the FASB issued ASU No. 2017-01, *Business Combinations (Topic 805) Clarifying the Definition of a Business* ("ASU 2017-01"). ASU 2017-01 clarifies the definition of a business by adding guidance to assist entities in evaluating whether transactions should be accounted for as acquisitions of assets or businesses. The definition of a business affects many areas of accounting including acquisitions, disposals, goodwill and consolidation. The ASU is effective for fiscal years beginning after December 15, 2018. Early adoption is permitted. The Company adopted this standard effective as of January 1, 2016 and applied it to its license and asset purchase agreements during the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited) (see Note 9).

In November 2016, the FASB issued ASU No. 2016-18, *Statement of Cash Flows (Topic 230): Restricted Cash* ("ASU 2016-18"), which requires that amounts generally described as restricted cash and restricted cash equivalents be included with cash and cash equivalents when reconciling the beginning-of-period and end-of-period total amounts shown on the statement of cash flows. ASU 2016-18 is effective for fiscal years beginning after December 15, 2017 and interim periods within those fiscal years and should be applied using a retrospective transition method to each period presented. Early adoption is permitted. The Company elected to early adopt ASU 2016-18 effective as of January 1, 2017 and has reflected the adoption retrospectively to all periods presented in its consolidated financial statements. As a result, the Company's consolidated statements of cash flows include restricted cash with cash and cash equivalents when reconciling the beginning-of-period and end-of-period total amounts shown on the such statements.

In August 2016, the FASB issued ASU No. 2016-15, Statement of Cash Flows (Topic 230): Classification of Certain Cash Receipts and Cash Payments ("ASU 2016-15"). This guidance addresses diversity in practice in how certain cash receipts and cash payments are presented in the statement of cash flows. The standard is effective for fiscal years beginning after December 15, 2017, including interim periods in those fiscal years, and early adoption is permitted. The adoption of ASU 2016-15 is required to be applied retrospectively. The Company adopted ASU 2017-09 as of

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

the required effective date of January 1, 2018, and the adoption did not have an impact on the Company's consolidated statement of cash flows.

In March 2016, the FASB issued ASU No. 2016-09, *Compensation — Stock Compensation (Topic 718): Improvements to Employee Share-Based Payment Accounting* ("ASU 2016-09"). ASU 2016-09 addresses several aspects of the accounting for share-based payment transactions, including the income tax consequences, classification of awards as either equity or liabilities, an option to recognize gross share compensation expense with actual forfeitures recognized as they occur, and classification on the statement of cash flows. Certain of these changes are required to be applied retrospectively, while other changes are required to be applied prospectively. The Company adopted ASU 2016-09 effective as of January 1, 2016 and elected prospectively to account for forfeitures as they occur rather than apply an estimated forfeiture rate to share-based compensation expense. The adoption of ASU 2016-09 did not have a material impact on the Company's financial position, results of operations or cash flows.

In November 2015, the FASB issued ASU No. 2015-17, *Income Taxes (Topic 740): Balance Sheet Classification of Deferred Taxes* ("ASU 2015-17"). ASU 2015-17 requires deferred tax liabilities and assets to be classified as non-current in the consolidated balance sheet. The amendment may be applied either prospectively to all deferred tax liabilities and assets or retrospectively to all periods presented. The Company adopted ASU 2015-17 effective as of January 1, 2016 and has reflected the adoption retrospectively to all periods presented in its consolidated financial statements. The adoption of ASU 2015-17 did not have a material impact on the Company's financial position, results of operations or cash flows.

In August 2014, the FASB issued ASU No. 2014-15, *Presentation of Financial Statements — Going Concern (Subtopic 205-40): Disclosure of Uncertainties about an Entity's Ability to Continue as a Going Concern* ("ASU 2014-15"). The amendments in this update explicitly require a company's management to assess if there is substantial doubt about an entity's ability to continue as a going concern within one year of the date of issuance of the entity's financial statements and to provide related footnote disclosures in certain circumstances. The Company adopted ASU 2014-15 effective as of January 1, 2016. This guidance relates to footnote disclosure only and its adoption had no impact on the Company's financial position, results of operations or cash flows.

In May 2014, the FASB issued ASU No. 2014-09, *Revenue from Contracts with Customers (Topic 606)* ("ASU 2014-09"), which supersedes existing revenue recognition guidance under GAAP. The standard's core principle is that a company will recognize revenue when it transfers promised goods or services to customers in an amount that reflects the consideration to which the company expects to be entitled in exchange for those goods or services. The standard defines a five-step process to achieve this principle, and will require companies to use more judgment and make more estimates than under the current guidance. The Company expects that these judgments and estimates will include identifying performance obligations in the customer contract, estimating the amount of variable consideration to include in the transaction price and allocating the transaction price to each separate performance obligation. ASU 2014-09 also requires additional disclosure about the nature, amount, timing and uncertainty of revenue and cash flows arising from customer contracts. In August 2015, the FASB issued ASU No. 2015-14, *Revenue from Contracts with Customers (Topic 606): Deferral of the Effective Date*, which delays the effective date of ASU

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

2. Summary of Significant Accounting Policies (Continued)

2014-09 such that the standard is effective for public entities for annual periods beginning after December 15, 2017 and for interim periods within those fiscal years. The FASB subsequently issued amendments to ASU 2014-09 that have the same effective date and transition date. The Company adopted ASU 2014-09 as of the required effective date of January 1, 2018 and the adoption did not have an impact on the Company's consolidated financial statements as the Company does not currently have any revenue-generating arrangements.

Recently Issued Accounting Pronouncements

In July 2017, the FASB issued ASU No. 2017-11, *Earnings Per Share (Topic 260), Distinguishing Liabilities from Equity (Topic 480), Derivatives and Hedging (Topic 815) I. Accounting for Certain Financial Instruments with Down Round Features II. Replacement of the Indefinite Deferral for Mandatorily Redeemable Financial Instruments of Certain Nonpublic Entities and Certain Mandatorily Redeemable Noncontrolling Interests with a Scope Exception ("ASU 2017-11").* Part I applies to entities that issue financial instruments such as warrants, convertible debt or convertible preferred stock that contain down-round features. Part II replaces the indefinite deferral for certain mandatorily redeemable noncontrolling interests and mandatorily redeemable financial instruments of nonpublic entities contained within ASC Topic 480 with a scope exception and does not impact the accounting for these mandatorily redeemable instruments. ASU 2017-11 is required to be adopted for annual periods beginning after December 15, 2018, including interim periods within those fiscal years. The Company is currently evaluating the impact that the adoption of ASU 2017-11 will have on its consolidated financial statements.

In February 2016, the FASB issued ASU No. 2016-02, *Leases (Topic 842)* ("ASU 2016-02"), which sets out the principles for the recognition, measurement, presentation and disclosure of leases for both parties to a contract (i.e., lessees and lessors). The new standard requires lessees to apply a dual approach, classifying leases as either finance or operating leases based on the principle of whether or not the lease is effectively a financed purchase by the lessee. This classification will determine whether lease expense is recognized based on an effective interest method or on a straight-line basis over the term of the lease, respectively. A lessee is also required to record a right-of-use asset and a lease liability for all leases with a term of greater than 12 months regardless of their classification. Leases with a term of 12 months or less will be accounted for similar to existing guidance for operating leases today. The guidance is effective for annual periods beginning after December 15, 2018 and for interim periods within those fiscal years, and early adoption is permitted. The Company is currently evaluating the impact that the adoption of ASU 2016-02 will have on its consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

3. Fair Value of Financial Assets and Liabilities

The following tables present information about the Company's financial assets and liabilities measured at fair value on a recurring basis and indicate the level of the fair value hierarchy used to determine such fair values:

	Fair Value Measurements as of December 31, 2016 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 105	$ —	$ —	$ 105
Cash equivalents — money market funds	550	—	—	550
Cash equivalents — U.S. Treasury notes	—	52,504	—	52,504
	$ 655	$ 52,504	$ —	$ 53,159

	Fair Value Measurements as of December 31, 2017 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 105	$ —	$ —	$ 105
Cash equivalents — money market funds	5,487	—	—	5,487
Cash equivalents — U.S. Treasury notes	—	14,995	—	14,995
	$ 5,592	$ 14,995	$ —	$ 20,587

	Fair Value Measurements as of March 31, 2018 Using:			
	Level 1	Level 2	Level 3	Total
	(unaudited)			
Assets:				
Restricted cash — money market funds (including non-current portion)	$ 315	$ —	$ —	$ 315
Cash equivalents — money market funds	110,267	—	—	110,267
Cash equivalents — U.S. Treasury notes	—	84,429	—	84,429
	$ 110,582	$ 84,429	$ —	$ 195,011

During the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited), there were no transfers between Level 1, Level 2 and Level 3.

The money market funds were valued using quoted prices in active markets, which represent a Level 1 measurement in the fair value hierarchy. The Company's cash equivalents as of December 31, 2016 and 2017 and March 31, 2018 (unaudited) also consisted of U.S. Treasury notes, which are not traded on a daily basis and, therefore, represent a Level 2 measurement in the fair value hierarchy at each period end.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

4. Property and Equipment, Net

Property and equipment, net consisted of the following:

	December 31,		March 31,
	2016	2017	2018
			(unaudited)
Furniture and fixtures	$ 14	$ 83	$ 6
Computer hardware and software	9	9	77
Vehicles	85	85	85
	108	177	168
Less: Accumulated depreciation	(24)	(52)	(42)
	$ 84	$ 125	$ 126

Depreciation expense for the years ended December 31, 2016 and 2017 was $22 and $28, respectively and $6 and $8 for the three months ended March 31, 2017 and 2018 (unaudited), respectively.

5. Accrued Expenses

Accrued expenses consisted of the following:

	December 31,		March 31,
	2016	2017	2018
			(unaudited)
Accrued employee compensation and benefits	$ 986	$ 1,570	$ 843
Accrued research and development expenses	979	3,905	3,742
Accrued legal and professional fees	122	688	1,599
Other	3	49	288
	$ 2,090	$ 6,212	$ 6,472

6. Convertible Preferred Shares

As of December 31, 2017, the Company's bye-laws, as amended and restated (the "Amended Bye-Laws"), designated 22,885,492 authorized shares to be issued as convertible preferred shares with a par value of $0.000273235 per share, of which 17,128,120 shares have been further designated as Series A convertible preferred shares (the "Series A preferred shares") and 5,757,372 shares have been further designated as Series B convertible preferred shares (the "Series B preferred shares"). As of March 31, 2018 (unaudited), the Company's bye-laws, as further amended and restated, designated 35,670,093 authorized shares to be issued as convertible preferred shares with a par value of $0.000273235 per share, of which 17,128,120 shares have been further designated as Series A preferred shares, 5,757,372 have been further designated as Series B preferred shares and 12,784,601 shares have been further designated as Series C convertible preferred shares (the "Series C preferred shares"). The holders of preferred shares have liquidation rights in the event of a deemed liquidation that, in certain situations, are not solely within

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

the control of the Company. Therefore, the Series A, Series B and Series C preferred shares (collectively, the "Preferred Shares") are classified outside of shareholders' equity (deficit).

In October 2015, the Company issued and sold 8,028,809 Series A preferred shares at a price of $4.6707 per share (the "Series A Original Issue Price") for proceeds of $37,398, net of issuance costs of $102.

In September 2016, the Company issued and sold an additional 9,099,311 Series A preferred shares at a price of $4.6707 per share for proceeds of $42,499, net of issuance costs of $1.

In March 2017, the Company issued and sold 5,757,372 Series B preferred shares at a price of $6.9475 per share (the "Series B Original Issue Price") for proceeds of $39,873, net of issuance costs of $127.

In February 2018, the Company issued and sold 12,784,601 Series C convertible preferred shares at a price of $15.6438 per share (the "Series C Original Issue Price") for proceeds of $190,822, net of issuance costs of $9,178.

As of each balance sheet date, the Preferred Shares consisted of the following:

	December 31, 2016				
	Preferred Shares Designated	Preferred Shares Issued and Outstanding	Carrying Value	Liquidation Preference	Common Shares Issuable Upon Conversion
Series A preferred shares	17,128,120	17,128,120	$ 79,897	$ 80,000	17,128,120

	December 31, 2017				
	Preferred Shares Designated	Preferred Shares Issued and Outstanding	Carrying Value	Liquidation Preference	Common Shares Issuable Upon Conversion
Series A preferred shares	17,128,120	17,128,120	$ 79,897	$ 80,000	17,128,120
Series B preferred shares	5,757,372	5,757,372	39,873	40,000	5,757,372
	22,885,492	22,885,492	$ 119,770	$ 120,000	22,885,492

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

	March 31, 2018 (unaudited)				
	Preferred Shares Designated	Preferred Shares Issued and Outstanding	Carrying Value	Liquidation Preference	Common Shares Issuable Upon Conversion
Series A preferred shares	17,128,120	17,128,120	$ 79,897	$ 80,000	17,128,120
Series B preferred shares	5,757,372	5,757,372	39,873	40,000	5,757,372
Series C preferred shares	12,784,601	12,784,601	190,822	200,000	12,784,601
	35,670,093	35,670,093	$ 310,592	$ 320,000	35,670,093

The holders of the Preferred Shares have the following rights and preferences:

Voting

The holders of Preferred Shares are entitled to vote, together with the holders of common shares, on all matters submitted to shareholders for a vote. The holders of Series A preferred shares are entitled to the number of votes per Series A preferred share equal to the number of whole Class B common shares into which the Series A preferred shares are convertible on the record date determining shareholders entitled to participate in such vote (which is ten votes for each Class B common share). The holders of Series B preferred shares are entitled to the number of votes per Series B preferred share equal to the number of whole Class A common shares into which the Series B preferred shares are convertible on the record date determining shareholders entitled to participate in such vote (which is one vote for each Class A common share). The holders of Series C preferred shares are entitled to the number of votes per Series C preferred share equal to the number of whole Class A common shares into which the Series C preferred shares are convertible on the record date determining shareholders entitled to participate in such vote (which is one vote for each Class A common share). Except as provided by law or by the other provisions of the Amended Bye-Laws, holders of Preferred Shares vote together with the holders of common shares as a single class.

The holders of Preferred Shares, voting together as a single class, are entitled to elect two directors of the Company. The holders of Preferred Shares, voting together with the holders of common shares as a single class, are entitled to elect the remaining directors of the Company, except for the one director that the holders of Class A common shares and Class B common shares, voting together as a single class, are entitled to elect.

Conversion

Each Series A preferred share shall be convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as is permitted by Bermuda law, into such number of fully paid and non-assessable Class B common shares as is determined by dividing the Series A Original Issue Price by the

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

Series A Conversion Price (as defined below) in effect at the time of conversion. Each Series B preferred share shall be convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as is permitted by Bermuda law, into such number of fully paid and non-assessable Class A common shares as is determined by dividing the Series B Original Issue Price by the Series B Conversion Price (as defined below) in effect at the time of conversion. Each Series C preferred share shall be convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as is permitted by Bermuda law, into such number of fully paid and non-assessable Class A common shares as is determined by dividing the Series C Original Issue Price by the Series C Conversion Price (as defined below) in effect at the time of conversion.

The Series A Original Issue Price and Series A Conversion Price were equal to $4.6707 as of December 31, 2016 and 2017 and March 31, 2018 (unaudited). The Series B Original Issue Price and Series B Conversion Price were equal to $6.9475 as of December 31, 2017 and March 31, 2018 (unaudited). The Series C Original Issue Price and Series C Conversion Price were equal to $15.6438 as of March 31, 2018 (unaudited). Such Series A, Series B and Series C Original Issue Prices and Series A, Series B and Series C Conversion Prices, and the rate at which each series of preferred shares may be converted into common shares, are subject to adjustment from time to time to reflect future share dividends, splits, combinations, recapitalizations and similar events. The Series A, Series B and Series C Conversion Prices are also subject to adjustments based on weighted-average anti-dilution provisions set forth in the Amended Bye-Laws in the event that additional securities are issued at a purchase price less than the Series A Conversion Price, the Series B Conversion Price or the Series C Conversion Price then in effect. As of December 31, 2016 and 2017 and March 31, 2018 (unaudited), each Series A preferred share was convertible into one Class B common share. As of December 31, 2017 and March 31, 2018 (unaudited), each Series B preferred share was convertible into one Class A common share. As of March 31, 2018 (unaudited), each Series C preferred share was convertible into one Class A common share.

Upon either (a) the closing of the sale of Class A common shares or Class B common shares to the public at a price of at least $15.6438 per share ($14.0120 prior to the issuance of the Series C preferred shares) (subject to appropriate adjustment in the event of any share dividend, share split, combination or other similar recapitalization with respect to the applicable class of common shares) in an initial public offering resulting in at least $100,000 of gross proceeds to the Company (a "Qualified IPO") or (b) the date and time, or the occurrence of an event, specified by vote or written consent of the holders of a majority of the outstanding Preferred Shares, voting together as a single class on an as-if-converted to Class A common shares basis, all outstanding Series A preferred shares shall automatically be converted, in such manner as is permitted pursuant to Bermuda law, into Class B common shares at the then effective conversion rate, and all outstanding Series B and Series C preferred shares shall automatically be converted, in such manner as is permitted pursuant to Bermuda law, into Class A common shares at the then effective conversion rate. In the event of a mandatory conversion of preferred shares as a result of a Qualified IPO, (i) holders of Series A preferred shares may elect to receive Class B1 common shares in lieu of Class B common shares and (ii) holders of Series B and Series C preferred shares may elect to receive Class A1 common shares in lieu of Class A common shares.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

Dividends

The holders of the Preferred Shares are entitled to receive noncumulative dividends when and if declared by Company's board of directors. The Company may not declare, pay or set aside any dividends on any other class or series of shares of the Company, other than dividends on common shares payable in common shares, unless the holders of the Preferred Share first receive, or simultaneously receive, a dividend on each outstanding Preferred Share equal to (A) in the case of a dividend on any class of common shares or any class or series that is convertible into common shares, that dividend per Preferred Share as would equal the product of (1) the dividend payable on each share of such class or series determined, if applicable, as if all shares of such class or series had been converted into common shares and (2) the number of common shares issuable upon conversion of a share the applicable series of Preferred Shares, or (B) in the case of a dividend on any class or series that is not convertible into common shares, at a rate per Preferred Share determined by (1) dividing the amount of the dividend payable on each share of such class or series of shares by the original issue price of such class or series (subject to appropriate adjustment in the event of any bonus share, share dividend, share split, combination of or other similar recapitalization with respect to such class or series) and (2) multiplying such fraction by an amount equal to the applicable Series A, Series B or Series C Original Issue Price. Through December 31, 2016 and 2017 and March 31, 2018 (unaudited), no cash dividends have been declared or paid.

Liquidation Preference

In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company or deemed liquidation event (as defined below), the holders of Preferred Shares then outstanding shall be entitled to be paid out of the assets of the Company available for distribution to its shareholders, on a *pari passu* basis, before any payment shall be made to the holders of common shares by reason of their ownership thereof, an amount per share equal to the greater of (i) one times the applicable Series A, Series B or Series C Original Issue Price, plus any dividends declared but unpaid thereon, and (ii) such amount per share as would have been payable had all Preferred Shares been converted into common shares immediately prior to such liquidation, dissolution, winding up or deemed liquidation event. Thereafter, the remaining assets of the Company available for distribution to its shareholders shall be distributed among the holders of common shares, pro rata based on the number of shares held by each such holder.

If upon any such liquidation, dissolution or winding up of the Company or deemed liquidation event, the assets of the Company available for distribution to its shareholders shall be insufficient to pay the holders of Preferred Shares the full amount to which they shall be entitled, the holders of Preferred Shares shall share ratably in any distribution of the assets available for distribution in proportion to the respective amounts which would otherwise be payable in respect of the shares held by such holders of Preferred Shares upon such distribution if all amounts payable on or with respect to such shares were paid in full.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

Unless a majority of the holders of the then outstanding Preferred Shares, on an as-if-converted to Class A common shares basis, elect otherwise, a deemed liquidation event shall include a merger or consolidation (other than one in which shareholders of the Company own a majority by voting power of the outstanding shares of the surviving or acquiring company or corporation) or a sale, lease, transfer, exclusive license or other disposition of all or substantially all of the assets of the Company.

Redemption

The Amended Bye-Laws do not provide redemption rights to the holders of Preferred Shares.

7. Common Shares

As of December 31, 2016, and 2017, the Amended Bye-Laws authorized the Company to issue 43,918,239 total shares with a par value of $0.000273235, of which 4,491,921 and 5,507,938 shares have been designated as Class A common shares as of December 31, 2016 and 2017, respectively, and 3,568,353 shares have been designated as Class B common shares as of December 31, 2016 and 2017. During the three months ended March 31, 2018 (unaudited), the Company amended and restated its Amended Bye-Laws to increase the total number of authorized shares to 44,746,463 shares, of which 5,507,938 shares have been designated as Class A common shares and 3,568,353 shares have been designated as Class B common shares as of March 31, 2018 (unaudited). The remaining 18,729,845, 11,956,456 and 79 shares that were not designated as common shares or Preferred Shares as of December 31, 2016 and 2017 and March 31, 2018 (unaudited), respectively, may be designated to any class at any time in the future by the Company's board of directors. No Class A1 common shares or Class B1 common shares were designated as of December 31, 2016 and 2017 and March 31, 2018 (unaudited). The rights of the holders of the Company's Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares are identical, except with respect to voting and conversion, as described below. The voting, dividend and liquidation rights of the holders of the Company's common shares are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares as set forth above.

Voting

Each Class A common share entitles the holder to one vote on all matters submitted to the shareholders for a vote. Each Class B common share entitles the holder to ten votes on all matters submitted to the shareholders for a vote. Holders of Class A1 common shares or Class B1 common shares have no voting rights. The holders of Class A and Class B common shares, voting together as a single class, are entitled to elect one director of the Company.

Dividends

Common shareholders are entitled to receive dividends, as may be declared by the board of directors. These dividends are subject to the preferential dividend rights of the holders of the Company's Preferred Shares. Through December 31, 2016 and 2017 and March 31, 2018 (unaudited), no cash dividends have been declared or paid.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

7. Common Shares (Continued)

Conversion

Each Class B common share shall automatically convert into one Class A common share upon certain transfers of such shares by the holder thereof (subject to certain exceptions). Each Class B common share shall be convertible, at the holder's election and at any time into one Class A common share or one Class B1 common share. Each Class A1 common share is convertible into one Class A common share at the holder's election. Each Class B1 common share is convertible into one Class A common share or one Class B common share at the holder's election.

There are no conversion rights associated with the Company's Class A common shares.

8. Share-Based Compensation

2015 Equity Incentive Plan

The Company's 2015 Equity Incentive Plan, as amended (the "2015 Plan"), provides for the Company to grant qualified incentive options, nonqualified options, share grants and other share-based awards to employees and non-employees to purchase the Company's Class A common shares.

The total number of common shares that may be issued under the 2015 Plan was 3,778,249 and 4,794,266 shares as of December 31, 2016 and 2017, respectively, of which 2,188,249 shares remained available for future grant as of December 31, 2016 and 1,664,893 shares remained available for future grant as of December 31, 2017. The total number of common shares that may be issued under the 2015 plan was 4,794,266 shares as of March 31, 2018 (unaudited), of which 81,193 shares remained available for future grant.

The exercise price for incentive options is determined by the board of directors. All incentive options granted to any person possessing less than 10% of the total combined voting power of all classes of shares may not have an exercise price of less than 100% of the fair market value of the Class A common shares on the grant date. All incentive options granted to any person possessing more than 10% of the total combined voting power of all classes of shares may not have an exercise price of less than 110% of the fair market value of the Class A common shares on the grant date. The option term for incentive awards may not be greater than 10 years. Incentive options granted to persons possessing more than 10% of the total combined voting power of all classes of shares may not have an option term of greater than five years. The vesting period for equity-based awards is determined by the board of directors, which is generally four to six years. For awards granted to employees and non-employees with four-year vesting terms, 25% of the option vests on the first anniversary of the grant date and the remaining shares vest equally each month for three years thereafter. For awards granted to employees with six-year vesting terms, 16% of the option vests on the first anniversary of the grant date and the remaining shares vest based on a predetermined vesting schedule for five years thereafter.

Shares that are expired, terminated, surrendered or canceled under the 2015 Plan without having been fully exercised will be available for future awards.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

8. Share-Based Compensation (Continued)

During the years ended December 31, 2016 and 2017, the Company granted options to purchase 316,866 and 1,545,045 Class A common shares, respectively, to employees and directors. During the three months ended March 31, 2017 and 2018 (unaudited), the Company granted options to purchase 37,512 and 1,652,321 Class A common shares, respectively, to employees and directors. The Company recorded share-based compensation expense for options granted to employees and directors of $354, $876, $110 and $536 during the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited), respectively.

During the years ended December 31, 2016 and 2017, the Company granted options to purchase 12,807 and 1,829 Class A common shares, respectively, to non-employees. During the three months ended March 31, 2017 (unaudited), the Company granted options to purchase 1,829 Class A common shares to non-employees. The Company did not grant any options to non-employees during the three months ended March 31, 2018 (unaudited). The Company recorded share-based compensation expense for options granted to non-employees of $14, $21, $4 and $22 during the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited), respectively.

Option Valuation

The assumptions that the Company used to determine the grant-date fair value of options granted to employees and directors were as follows, presented on a weighted-average basis:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
				(unaudited)
Risk-free interest rate	1.45%	1.99%	2.17%	2.71%
Expected term (in years)	6.25	6.25	6.25	6.56
Expected volatility	70.75%	74.18%	74.03%	75.25%
Expected dividend yield	0%	0%	0%	0%

The assumptions that the Company used to determine the fair value of options granted to non-employees were as follows, presented on a weighted-average basis:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
				(unaudited)
Risk-free interest rate	1.94%	2.49%	2.49%	2.78%
Expected term (in years)	10.00	10.00	10.00	7.89
Expected volatility	65.85%	78.28%	78.28%	73.96%
Expected dividend yield	0%	0%	0%	0%

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

8. Share-Based Compensation (Continued)

Options

Through December 31, 2017 and March 31, 2018 (unaudited), all options granted by the Company under the 2015 Plan were for the purchase of Class A common shares. The following table summarizes option activity under the 2015 Plan for the year ended December 31, 2017 and the three months ended March 31, 2018 (unaudited):

	Number of Shares	Weighted Average Exercise Price	Weighted Average Remaining Contractual Term (in years)	Aggregate Intrinsic Value
Outstanding as of December 31, 2016	1,583,691	$ 1.64	9.10	$ 356
Granted	1,546,874	3.90		
Exercised	—	—		
Forfeited	(7,501)	3.75		
Outstanding as of December 31, 2017	3,123,064	$ 2.75	8.82	$ 6,010
Granted	1,652,321	10.36		
Exercised	(4,574)	3.80		
Forfeited	(68,621)	3.80		
Outstanding as of March 31, 2018 (unaudited)	4,702,190	$ 5.41	9.03	$ 23,280
Options exercisable as of December 31, 2017	843,454	$ 1.62	8.07	$ 2,580
Options exercisable as of March 31, 2018 (unaudited)	955,119	$ 1.64	7.82	$ 8,333
Options unvested as of December 31, 2017	2,279,610	$ 3.17	9.09	$ 3,431
Options unvested as of March 31, 2018 (unaudited)	3,747,071	$ 6.37	9.34	$ 14,947

The aggregate intrinsic value of options is calculated as the difference between the exercise price of the options and the fair value of the Company's common shares for those options that had exercise prices lower than the fair value of the Company's common shares.

During the year ended December 31, 2016, an option holder exercised 6,309 options for Class A common shares with an intrinsic value of $2 for total cash proceeds of $10. There were no options exercised during the year ended December 31, 2017. During the three months ended March 31, 2018 (unaudited), an option holder exercised 4,574 options for Class A common shares with an instrinsic value of $30 for total cash proceeds of $17.

The weighted-average grant-date fair value per share of options granted during the years ended December 31, 2016 and 2017 was $1.16 and $2.57, respectively. The weighted-average grant-date fair value per share of options granted during the three months ended March 31, 2017 and 2018 (unaudited) was $2.54 and $7.18, respectively.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

8. Share-Based Compensation (Continued)

The total fair value of options vested during the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited) was $402, $445, $89 and $126, respectively.

Restricted Shares

Under terms of the Class A and Class B restricted share agreements covering the Class A and Class B common shares, restricted common shares are subject to a vesting schedule. The restricted shares vest over a four-year period during which time the Company has the right to repurchase up to all unvested shares at the amount paid if the relationship between the recipient and the Company ceases. Subject to the continued employment (or other engagement of the recipient by the Company as described in the restricted share agreements), all of the restricted common shares become fully vested within four years of the date of issuance.

The following table summarizes restricted share activity for the year ended December 31, 2017 and the three months ended March 31, 2018 (unaudited):

	Class A		Class B	
	Number of Shares	Weighted Average Fair Value at Issuance	Number of Shares	Weighted Average Fair Value at Issuance
Unvested restricted shares outstanding as of December 31, 2016	490,646	$ 0.000273235	2,527,583	$ 0.000273235
Granted	—	—	—	—
Vested	(178,417)	0.000273235	(892,088)	0.000273235
Unvested restricted shares outstanding as of December 31, 2017	312,229	$ 0.000273235	1,635,495	$ 0.000273235
Granted	—	—	—	—
Vested	(44,604)	0.000273235	(223,022)	0.000273235
Unvested restricted shares outstanding as of March 31, 2018 (unaudited)	267,625	$ 0.000273235	1,412,473	$ 0.000273235

The aggregate fair value of restricted shares that vested during the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited) was $2,348, $3,973, $670 and $1,757, respectively.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

8. Share-Based Compensation (Continued)

Share-Based Compensation

Share-based compensation expense was classified in the consolidated statements of operations and comprehensive loss as follows:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
				(unaudited)
Research and development expenses	$ 59	$ 324	$ 30	$ 202
General and administrative expenses	309	573	84	356
	$ 368	$ 897	$ 114	$ 558

As of December 31, 2017 and March 31, 2018 (unaudited), total unrecognized compensation cost related to the unvested share-based awards was $4,280 and $16,226, respectively, which is expected to be recognized over a weighted average remaining period of 3.22 years and 4.58 years, respectively.

9. License and Acquisition Agreements

Biogen Asset Purchase Agreement

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716 and other antibodies covered by certain patent rights, including patents and other intellectual property rights, clinical data, know-how, and clinical drug supply. In addition, Biogen granted to the Company a non-exclusive, sublicensable, worldwide license to certain background patent rights related to the KPL-716 program. The Company is obligated to use commercially reasonable efforts to develop and commercialize such acquired products.

In exchange for these rights, the Company made an upfront payment to Biogen of $11,500 and a technology transfer payment of $500. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment and technology transfer payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

Under the Biogen Agreement, the Company is obligated to make milestone payments to Biogen of up to $179,000 upon the achievement of specified clinical and regulatory milestones in multiple indications in various territories. During the year ended December 31, 2017, the Company made a milestone payment of $4,000 associated with the achievement of a specified clinical milestone event. Additionally, the Company could be obligated to make up to an aggregate of up to $150,000 of payments upon the achievement of specified annual net sales milestones and to pay

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

The Company also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to the KPL-716 program. Under these retained contracts, the Company paid a one-time upfront sublicense fee of $150 and is obligated to pay insignificant annual maintenance fees as well as clinical and regulatory milestone payments of up to an aggregate of $1,575. During the year ended December 31, 2017 the Company paid $75 upon the achievement of certain milestones in connection with the retained contracts. There were no milestone payments made during the three months ended March 31, 2018 (unaudited).

The Biogen Agreement will terminate upon the expiration of all payment obligations with respect to the last product in all countries in the territory. The Company has the right to terminate the agreement with 90 days' prior written notice. Both parties may terminate by mutual written consent or in the event of material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches).

During the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited), the Company recorded research and development expense in connection with the Biogen Agreement of $12,100, $4,169, $61 and $11, respectively.

Novo Nordisk License Agreement

In August 2017, the Company entered into a license agreement (the "Novo Nordisk Agreement") with Novo Nordisk A/S ("Novo Nordisk"), pursuant to which the Company has been granted an exclusive, sublicensable, worldwide license under certain intellectual property rights controlled by Novo Nordisk to make, use, develop and commercialize KPL-045 for all indications. The Company is obligated to use commercially reasonable efforts to develop and commercialize such licensed products.

In consideration for the license, the Company made an upfront payment of $1,500 to Novo Nordisk. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

Under the Novo Nordisk Agreement, the Company is also required to make a payment of $150 upon completion of the technology transfer by Novo Nordisk. The technology was transferred during the three months ended March 31, 2018 and, as a result, this payment was made and is recorded in the Company's statement of operations for the three months ended March 31, 2018 (unaudited). In addition, the Company is obligated to make milestone payments upon the achievement of specified clinical, regulatory and initial sales milestones and upon the achievement of annual net sales thresholds, including a payment of $1,000 upon the earlier to occur of a specified regulatory milestone and January 2020, unless the Novo Nordisk Agreement is earlier terminated by either party. As of December 31, 2017 and March 31, 2018 (unaudited), the Company determined that the payment related to the milestone was not probable and, therefore, no

9. License and Acquisition Agreements (Continued)

amount was recorded in the Company's consolidated statement of operations and comprehensive loss during the year ended December 31, 2017 and the three months ended March 31, 2018 (unaudited). The Company has also agreed to pay royalties on annual net sales of products licensed under the agreement.

Under the Novo Nordisk Agreement, the Company is solely responsible for all development, regulatory and commercial activities and costs. The Company is also responsible for costs related to filing, prosecuting and maintaining the licensed patent rights.

The Novo Nordisk Agreement will terminate upon expiration of the last-to-expire royalty term for any licensed product in the territories, as defined in the agreement. Either party may terminate the agreement upon the other party's insolvency or bankruptcy or for uncured material breach of the agreement by the other party. Novo Nordisk has the right to terminate the agreement if the Company challenges any of the licensed patent rights. The Company may also terminate the agreement for any reason upon prior written notice to Novo Nordisk.

During the year ended December 31, 2017 and the three months ended March 31, 2018 (unaudited), the Company recorded research and development expense of $1,500 and $150, respectively, in connection with the Novo Nordisk Agreement.

Primatope Stock Purchase Option Agreement

In September 2017, the Company entered into a stock purchase option agreement (the "Primatope Agreement") with Primatope Therapeutics, Inc. ("Primatope"), pursuant to which the Company has been granted a license to certain intellectual property rights controlled by Primatope to research, develop, and manufacture the pre-clinical antibody, KPL-404.

The agreement provides the Company with an exclusive call option to purchase 100% of the capital stock of Primatope. Upon execution of the agreement, the Company made $500 in upfront payments for the initial option period through April 2018 (the "Initial Option Period"). The Primatope Agreement allows up to three extensions of the Initial Option Period through January 2019 (including the initial option period, the "Option Period") for total extension payments of up to $800. In April 2018, the Company made a payment of $250 to extend the Option Period for an additional three months to June 2018. During the Option Period, the Company may conduct research and pre-clinical work to assess the viability of the asset.

If the call option is exercised, the Company will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10,000 as well as potential milestone payments of up to $10,000. The upfront payment and the milestone payments may be paid in a combination of cash and issuance of the Company's Class A common shares.

The Company has determined that the call option represents a variable interest in Primatope and that Primatope is a VIE. However, as the Company has no ability to control the board of directors or direct the ongoing activities of Primatope, the Company does not have power over the activities that most significantly impact Primatope's economic performance and is not the primary beneficiary of Primatope. As a result, the Company does not consolidate the assets, liabilities, and results of operations of Primatope.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

Either party may terminate the Primatope Agreement for uncured material breach of the agreement by the other party or by mutual written consent.

During the year ended December 31, 2017, the Company recorded research and development expense of $500 in connection with the Primatope Agreement. The Company did not incur any research and development expense in connection with the Primatope Agreement during the three months ended March 31, 2018 (unaudited).

Regeneron License Agreement

In September 2017, the Company entered into a license agreement (the "Regeneron Agreement") with Regeneron Pharmaceuticals, Inc. ("Regeneron"), pursuant to which the Company has been granted an exclusive, sublicensable license under certain intellectual property rights controlled by Regeneron to develop and commercialize rilonacept in certain fields and territories. The Company is obligated to use commercially reasonable efforts to develop and commercialize such licensed products.

In exchange for these rights, the Company made an upfront payment of $5,000. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

Under the Regeneron Agreement, the Company is also obligated to make payments to Regeneron of up to an aggregate of $27,500 upon the achievement of specified regulatory milestones. Upon commercialization of the licensed products, the parties will share profits equally, after deducting certain commercialization expenses subject to specified limits.

Under the Regeneron Agreement, the Company is solely responsible for all development and commercialization activities and costs in its respective territory. The Company is also responsible for costs related to the filing, prosecution and maintenance of certain licensed patent rights.

The parties also entered into a clinical supply agreement under which Regeneron agreed to manufacture the developed product during the clinical phase. During the year ended December 31, 2017 and the three months ended March 31, 2018 (unaudited), the Company recognized research and development expense of $208 and $376, respectively related to the purchase of drug materials under this agreement. As of December 31, 2017 and March 31, 2018 (unaudited), the Company has non-cancelable purchase commitments under the clinical supply agreement (see Note 12).

The Regeneron Agreement will expire when the Company is no longer developing or commercializing any licensed product under the Regeneron Agreement. Either party may terminate the agreement upon the other party's insolvency or bankruptcy or for material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches). Regeneron has the right to terminate the agreement if the Company suspends its development or commercialization activities for a consecutive 12 month period or does not grant a sublicense to a third-party to perform such activities, or if the Company challenges any of the licensed patent rights. The Company may terminate the agreement at any time that is 18 months

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

after the effective date of the agreement with 180 days' written notice or with one years' written notice if we terminate the agreement following U.S. marketing approval of a rilonacept product developed by the Company. The Company may also terminate the agreement with three month's written notice if the products are determined to have certain safety concerns.

During the year ended December 31, 2017 and the three months ended March 31, 2018 (unaudited), the Company recorded research and development expense of $5,208 and $376, respectively, in connection with the agreements with Regeneron.

MedImmune License Agreement

In December 2017, the Company entered into a license agreement (the "MedImmune Agreement") with MedImmune, Limited ("MedImmune"), pursuant to which MedImmune granted the Company an exclusive, sublicensable, worldwide license to certain intellectual property rights to make, use, develop and commercialize mavrilimumab. Under the MedImmune Agreement, the Company also acquired reference rights to relevant manufacturing and regulatory documents and MedImmune's existing supply of mavrilimumab drug substance and product. The Company is obligated use commercially reasonable efforts to develop and commercialize the licensed products.

In exchange for these rights, the Company made an upfront payment of $8,000. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use. In addition, the Company is obligated to make clinical, regulatory and initial sales milestone payments of up to $72,500 in aggregate for the first two indications, including a milestone payment of $10,000 upon the earlier to occur of a specified regulatory milestone and December 31, 2018, unless the MedImmune Agreement is earlier terminated by either party. As of December 31, 2017, the Company determined that the payment related to this milestone was probable and, therefore, recognized research and development expense and an accrued milestone of $10,000 during the year ended December 31, 2017. In addition, the Company is obligated to make clinical and regulatory milestone payments of up to $15,000 in the aggregate for each subsequent indication. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

The Company is solely responsible for all development, manufacturing, and commercial activities and costs of the licensed products, including clinical studies or other tests necessary to support the use of a licensed product. The Company is also responsible for costs related to the filing, prosecution and maintenance of the licensed patent rights.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

The MedImmune Agreement will expire upon the expiration of the royalty term in the last country for the last indication, as defined in the agreement. Either party may terminate the agreement upon the other party's insolvency or bankruptcy or for material breach of the agreement by the other party that remains uncured for 90 days. MedImmune has the right to terminate the agreement if the Company challenges any of the licensed patent rights. The Company may terminate the agreement at any time upon 90 days' prior written notice.

During the year ended December 31, 2017, the Company recorded research and development expense of $18,000 in connection with the MedImmune Agreement. The Company did not incur any research and development expense in connection with the MedImmune Agreement during the three months ended March 31, 2018 (unaudited).

10. Income Taxes

As a company incorporated in Bermuda, the Company is principally subject to taxation in Bermuda. Under the current laws of Bermuda, tax on a company's income is assessed at a zero percent tax rate. As a result, the Company has not recorded any income tax benefits from its losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards will be available to the Company for those losses.

In August 2015, the Company entered into agreements with its wholly owned subsidiary, Kiniksa US, under which Kiniksa US provides management and research and development services to the Company for which the Company pays costs plus a service fee. Kiniksa US is subject to tax for federal and state tax purposes. On December 22, 2017, the United States enacted new tax reform ("Tax Cuts and Jobs Act"). The Tax Cuts and Jobs Act contains provisions with separate effective dates but is generally effective for taxable years beginning after December 31, 2017. Beginning with the year ending December 31, 2018, the corporate statutory rates on U.S. earnings will be reduced from a top marginal rate of 35% to a flat rate of 21%. The impact of the future rate reduction resulted in a provision for income taxes of $69 for the year ended December 31, 2017 relating to the revaluation of the Company's net deferred tax assets.

Income (loss) before provision for income taxes consisted of the following:

	Year Ended December 31,	
	2016	**2017**
Bermuda	$ (24,254)	$ (65,391)
Foreign (U.S.)	317	520
	$ (23,937)	$ (64,871)

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

10. Income Taxes (Continued)

The components of the Company's income tax provision for the years ended December 31, 2016 and 2017 are as follows:

	Year Ended December 31,	
	2016	2017
Current income tax provision:		
Bermuda	$ —	$ —
U.S. federal	78	184
U.S. state	4	15
Total current income tax provision	82	199
Deferred income tax provision (benefit):		
Bermuda	—	—
U.S. federal	(26)	(87)
U.S. state	(20)	(110)
Total deferred income tax provision (benefit)	(46)	(197)
Total provision for income taxes	$ 36	$ 2

A reconciliation of the Bermuda statutory income tax rate of 0% to the Company's effective income tax rate is as follows:

	Year Ended December 31,	
	2016	2017
Bermuda statutory income tax rate	—%	—%
Foreign (U.S.) tax rate differential	(0.5)	(0.4)
Research and development tax credits	0.5	0.5
2017 Tax Cuts and Jobs Act	—	(0.1)
Effective income tax rate	—%	—%

The Company recorded immaterial tax benefits in the three months ended March 31, 2017 and 2018 (unaudited). The Company expects an immaterial level of tax benefit in 2018, comprised of current tax expense related to the Company's operations in the U.S. which is more than offset by federal and state research and development tax credits.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

10. Income Taxes (Continued)

Net deferred tax assets consisted of the following:

	December 31,	
	2016	2017
Research and development tax credit carryforwards	$ 18	$ 90
Depreciation and amortization	(4)	(14)
Accrued expenses and other	37	189
Total deferred tax assets	51	265
Valuation allowance	(10)	(27)
Net deferred tax assets	$ 41	$ 238

As of December 31, 2017, the Company had state research and development tax credit carryforwards of approximately $113, available to reduce future tax liabilities, which begin to expire in 2031 through 2032. As of March 31, 2018 (unaudited), the amount of the Company's deferred tax assets did not change materially from the amount as of December 31, 2017.

As required by ASC 740, the Company has evaluated the positive and negative evidence bearing upon its ability to realize the deferred tax assets. In order to utilize state research and development tax credits, the Company will need taxable income in the jurisdiction of where the credit was generated. The Company currently has no taxable income in certain state jurisdictions and thus management has determined that it is more likely than not that the Company will not recognize the benefits of state research and development tax credits generated in those jurisdictions, and as a result, a valuation allowance of $10 and $27 has been established at December 31, 2016 and 2017, respectively. The remaining deferred tax assets will be fully utilized in the United States based on future income generated under the cost-plus arrangement in place.

Utilization of the state research and development tax credits may be subject to substantial annual limitation under Section 382 of the Internal Revenue Code of 1986 due to ownership changes that could occur in the future. These ownership changes may limit the amount of carryforwards that can be utilized annually to offset future taxable income. In general, an ownership change, as defined by Section 382, results from transactions increasing the ownership of certain shareholders or public groups in the stock of a corporation by more than 50% over a three-year period.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

10. Income Taxes (Continued)

Changes in the valuation allowance for deferred tax assets during the years ended December 31, 2016 and 2017 were due primarily to an increase in state research and development tax credits and were as follows:

	Year Ended December 31,	
	2016	2017
Valuation allowance at beginning of year	$ (1)	$ (10)
Increases recorded to income tax provision	(9)	(17)
Valuation allowance at end of year	$ (10)	$ (27)

The Company has not recorded any amounts for unrecognized tax benefits as of December 31, 2016 or 2017. The Company's policy is to record interest and penalties related to income taxes as part of its income tax provision. As of December 31, 2016 and 2017, the Company had no accrued interest or penalties related to uncertain tax positions and no amounts had been recognized in the Company's consolidated statements of operations and comprehensive loss.

The Company files income tax returns in the United States and certain state jurisdictions. Kiniksa US's federal and state income tax returns are subject to tax examinations for the tax years ended December 31, 2013 and subsequent years. To the extent the Company has tax attribute carryforwards, the tax years in which the attribute was generated may still be adjusted upon examination by the Internal Revenue Service and state tax authorities to the extent utilized in a future period. There are currently no income tax examinations pending.

11. Net Loss per Share and Unaudited Pro Forma Net Loss per Share

Net Loss per Share

The rights, including the liquidation and dividend rights, of the holders of Class A and Class B common shares are identical, except with respect to voting rights. As the liquidation and dividend rights are identical, losses are allocated on a proportionate basis and the resulting net loss per share attributed to common shareholders will, therefore, be the same for both Class A and Class B common shares on an individual or combined basis.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

11. Net Loss per Share and Unaudited Pro Forma Net Loss per Share (Continued)

Basic and diluted net loss per share attributable to common shareholders was calculated as follows:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
			(unaudited)	
Numerator:				
Net loss attributable to common shareholders	$ (23,973)	$ (64,873)	$ (4,941)	$ (15,982)
Denominator:				
Weighted average common shares outstanding — basic and diluted	261,695	1,809,751	1,405,400	2,478,903
Net loss per share attributable to common shareholders — basic and diluted	$ (91.61)	$ (35.85)	$ (3.52)	$ (6.45)

The Company's unvested restricted common shares have been excluded from the computation of basic net loss per share attributable to common shareholders.

The Company's potentially dilutive securities, which include options, unvested restricted shares and convertible preferred shares, have been excluded from the computation of diluted net loss per share attributable to common shareholders as the effect would be to reduce the net loss per share attributable to common shareholders. Therefore, the weighted average number of common shares outstanding used to calculate both basic and diluted net loss per share attributable to common shareholders is the same. The Company excluded the following potential common shares, presented based on amounts outstanding at each period end, from the computation of diluted net loss per share attributable to common shareholders for the periods indicated because including them would have had an anti-dilutive effect:

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
			(unaudited)	
Options to purchase common shares	1,583,691	3,123,064	1,622,850	4,702,190
Unvested restricted shares	3,018,229	1,947,724	2,750,603	1,680,098
Convertible preferred shares (as converted to common shares)	17,128,120	22,885,492	22,885,492	35,670,093
	21,730,040	27,956,280	27,258,945	42,052,381

Unaudited Pro Forma Net Loss per Share

The unaudited pro forma basic and diluted net loss per share attributable to common shareholders for the year ended December 31, 2017 and the three months ended March 31, 2018

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

11. Net Loss per Share and Unaudited Pro Forma Net Loss per Share (Continued)

have been prepared to give effect to adjustments arising upon the closing of a qualified initial public offering.

The unaudited pro forma basic and diluted weighted average common shares outstanding used in the calculation of unaudited pro forma basic and diluted net loss per share attributable to common shareholders for the year ended December 31, 2017 and the three months ended March 31, 2018 have been prepared to give effect, upon a qualified initial public offering, to the automatic conversion of (i) all outstanding Series A preferred shares into Class B common shares and Class B1 common shares and (ii) all outstanding Series B and Series C preferred shares into Class A common shares and Class A1 common shares as if the proposed initial public offering had occurred on the later of January 1, 2017 or the issuance date of the Preferred Shares.

The rights, including the liquidation and dividend rights, of the holders of Class A, Class A1, Class B and Class B1 common shares are identical, except with respect to voting rights. As the liquidation and dividend rights are identical, losses are allocated on a proportionate basis and the resulting unaudited pro forma net loss per share attributed to common shareholders will, therefore, be the same for both Class A, Class A1, Class B and Class B1 common shares on an individual or combined basis.

Unaudited pro forma basic and diluted net loss per share attributable to common shareholders was calculated as follows:

	Year Ended December 31, 2017	Three Months Ended March 21, 2018
	(unaudited)	
Numerator:		
Net loss attributable to common shareholders	$ (64,873)	$ (15,982)
Denominator:		
Weighted average common shares outstanding — basic and diluted	1,809,751	2,478,903
Pro forma adjustment to reflect assumed automatic conversion of Preferred Shares upon the closing of the proposed initial public offering	21,828,659	29,988,048
Pro forma weighted average common shares outstanding — basic and diluted	23,638,410	32,466,951
Pro forma net loss per share attributable to common shareholders — basic and diluted	$ (2.74)	$ (0.49)

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

12. Commitments and Contingencies

Lease Agreements

On July 24, 2015, Kiniksa US entered into an operating lease in Wellesley Hills, Massachusetts for office space that comprises the headquarters for Kiniksa US. In March 2016, effective August 1, 2016, Kiniksa US entered into an expansion and extension on its lease, which expanded its leased space to a total of 10,800 square feet. On March 31, 2017, Kiniksa US renewed this lease and extended the lease term to August 2018. Monthly lease payments, inclusive of base rent and ancillary charges, total $27. As of December 31, 2017 future minimum lease payments under non-cancelable operating lease commitments, which are all due during the year ending December 31, 2018, totaled $270.

On March 13, 2018, Kiniksa US entered into an operating lease in Lexington, Massachusetts for office and laboratory space that comprises the new headquarters for Kiniksa US. The lease expires on July 31, 2021. Upon execution of the lease, the Company made a prepayment of $67 for August 2018 base rent. Monthly lease payments begin in September 2018 and include base rent of $67 as well as, ancillary charges such as the share of operating expenses and real estate taxes.

The following table summarizes the future minimum lease payments under non-cancelable operating lease commitments, for the Lexington office, as of March 31, 2018 (unaudited):

Year Ending December 31,	
2018	$ 267
2019	802
2020	802
2021	468
	$ 2,339

The Company recognizes rent expense on a straight-line basis over the respective lease period and has recorded deferred rent for rent expense incurred but not yet paid. The Company recorded rent expense of $286, $402, $100 and $130 during the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited), respectively.

License Agreements

The Company has entered into license agreements with various parties under which it is obligated to make contingent and non-contingent payments (see Note 9).

Manufacturing Commitments

During the year ended December 31, 2017 and three months ended March 31, 2018 (unaudited), the Company entered into agreements with several contract manufacturing organizations to provide pre-clinical and clinical trial materials. As of December 31, 2017 and March 31, 2018 (unaudited), the Company had non-cancelable purchase commitments under these agreements totaling $7,766 and $5,870, respectively, which are all due during the year ending December 31, 2018.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

12. Commitments and Contingencies (Continued)

Indemnification Agreements

In the ordinary course of business, the Company may provide indemnification of varying scope and terms to vendors, lessors, business partners and other parties with respect to certain matters including, but not limited to, losses arising out of breach of such agreements or from intellectual property infringement claims made by third parties. In addition, the Company has entered into indemnification agreements with members of its board of directors and executive officers that will require the Company, among other things, to indemnify them against certain liabilities that may arise by reason of their status or service as directors or officers. The maximum potential amount of future payments the Company could be required to make under these indemnification agreements is, in many cases, unlimited. To date, the Company has not incurred any material costs as a result of such indemnifications. The Company does not believe that the outcome of any claims under indemnification arrangements will have a material effect on its financial position, results of operations or cash flows, and it has not accrued any liabilities related to such obligations in its consolidated financial statements as of December 31, 2016 or 2017.

Legal Proceedings

The Company is not a party to any litigation and does not have contingency reserves established for any litigation liabilities.

13. Benefit Plans

The Company has established a defined-contribution savings plan under Section 401(k) of the Internal Revenue Code. This plan covers substantially all employees who meet minimum age and service requirements and allows participants to defer a portion of their annual compensation on a pre-tax basis. The Company provides matching contributions of 100% of the first 3% of each participant's salary contributed, plus 50% for each of the next 2% contributed. Employees are immediately and fully vested in their own contributions and the Company's match. During the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018 (unaudited), the Company contributed $143, $264, $75 and $115, respectively, to the plan.

14. Subsequent Events

For its consolidated financial statements as of December 31, 2017 and for the year then ended, the Company evaluated subsequent events through February 27, 2018, the date on which those financial statements were issued, and, with respect to the reverse share split described below, through May 14, 2018.

Reverse Share Split

On May 11, 2018, the Company effected a 1-for-2.73235 reverse share split of its authorized, designated, issued and outstanding common shares and Preferred Shares. Accordingly, all share and per share amounts for all periods presented in the accompanying consolidated financial statements and notes thereto have been adjusted retroactively, where applicable, to reflect this reverse share split.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

15. Subsequent Events (Unaudited)

For its interim consolidated financial statements as of March 31, 2018 and for the three months then ended, the Company evaluated subsequent events through April 27, 2018, the date on which those financial statements were issued, and, with respect to the reverse share split described above, through May 14, 2018.

2018 Incentive Award Plan

On May 11, 2018, the Company's board of directors and shareholders approved the 2018 Incentive Award Plan (the "2018 Plan"), which will become effective upon the date of the effectiveness of the registration statement for the Company's initial public offering. The 2018 Plan provides for the grant of incentive options, nonqualified options, share appreciation rights, restricted shares, dividend equivalents, restricted share units and other share- or cash-based awards. A total of 4,466,500 Class A common shares were initially reserved for issuance under the 2018 Plan. The number of Class A common shares that may be issued under the 2018 Plan will automatically increase on each January 1, beginning with the fiscal year ending December 31, 2019 and continuing for each fiscal year until, and including, the fiscal year ending December 31, 2028, equal to the lesser of (i) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (ii) a smaller number of Class A common shares determined by the Company's board of directors. No more than 27,915,000 Class A common shares may be issued under the 2018 Plan upon the exercise of incentive options. The Class A common shares underlying any awards that expire, lapse unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited under the 2018 Plan or the 2015 Plan will be added back to the Class A common shares available for issuance under the 2018 Plan.

2018 Employee Share Purchase Plan

On May 11, 2018, the Company's board of directors and shareholders approved the 2018 Employee Share Purchase Plan (the "2018 ESPP"), which will become effective upon the date of the effectiveness of the registration statement for the Company's initial public offering. A total of 670,000 Class A common shares were initially reserved for issuance under the 2018 ESPP. The number of Class A common shares that may be issued under the 2018 ESPP will automatically increase on each January 1, beginning with the fiscal year ending December 31, 2019 and continuing for each fiscal year until, and including, the fiscal year ending December 31, 2028, equal to the lesser of (i) 1% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (ii) a smaller number of Class A common shares determined by the Company's board of directors, provided that no more than 6,420,000 Class A common shares may be issued under the 2018 ESPP.

7,000,000 Class A Common Shares

Goldman, Sachs & Co. LLC
J.P. Morgan
JMP Securities
Wedbush PacGrow

Through and including , 2018 (the 25th day after the date of this prospectus), all dealers effecting transactions in these securities, whether or not participating in this offering, may be required to deliver a prospectus. This is in addition to a dealer's obligation to deliver a prospectus when acting as an underwriter and with respect to an unsold allotment or subscription.

Part II

INFORMATION NOT REQUIRED IN PROSPECTUS

Item 13. ***Other Expenses of Issuance and Distribution.***

The following table indicates the expenses to be incurred in connection with this offering described in this registration statement, other than underwriting discounts and commissions, all of which will be paid by us. All amounts are estimated except the SEC registration fee, the Financial Industry Regulatory Authority, Inc., or FINRA, filing fee and The Nasdaq Global Market fee.

	Amount
SEC Registration fee	$ 19,043
FINRA filing fee	23,442
The Nasdaq Global Market initial listing fee	125,000
Accountants' fees and expenses	1,000,000
Legal fees and expenses	2,270,000
Blue Sky fees and expenses	10,000
Transfer Agent's fees and expenses	5,000
Printing and engraving expenses	200,000
Miscellaneous	147,515
Total expenses	$ 3,800,000

Item 14. ***Indemnification of Directors and Officers.***

Section 98 of the Companies Act provides generally that a Bermuda company may indemnify its directors, officers and auditors against any liability which by virtue of any rule of law would otherwise be imposed on them in respect of any negligence, default, breach of duty or breach of trust, except in cases where such liability arises from fraud or dishonesty of which such director, officer or auditor may be guilty in relation to the company. Section 98 further provides that a Bermuda company may indemnify its directors, officers and auditors against any liability incurred by them in defending any proceedings, whether civil or criminal, in which judgment is awarded in their favor or in which they are acquitted or granted relief by the Supreme Court of Bermuda pursuant to section 281 of the Companies Act.

We have adopted provisions in our bye-laws that provide that we shall indemnify our officers and directors in respect of their actions and omissions, except in respect of their fraud or dishonesty. Our bye-laws provide that the shareholders waive all claims or rights of action that they might have, individually or in right of the Company, against any of the Company's directors or officers for any act or failure to act in the performance of such director's or officer's duties, except in respect of any fraud or dishonesty of such director or officer. Section 98A of the Companies Act permits us to purchase and maintain insurance for the benefit of any officer or director in respect of any loss or liability attaching to him in respect of any negligence, default, breach of duty or breach of trust, whether or not the Company may otherwise indemnify such officer or director. We will maintain a general liability insurance policy that covers certain liabilities of directors and officers of our Company arising out of claims based on acts or omissions in their capacities as directors or officers.

We have entered into indemnification agreements with each of our directors and officers. These indemnification agreements may require us, among other things, to indemnify our directors and officers for some expenses, including attorneys' fees, judgments, fines and settlement amounts incurred by a director or officer in any action or proceeding arising out of his or her service as one

of our directors or officers, or any other company or enterprise to which the person provides services at our request.

In any underwriting agreement we enter into in connection with the sale of Class A common shares being registered hereby, the underwriters will agree to indemnify, under certain conditions, us, our directors, our officers and persons who control us within the meaning of the Securities Act against certain liabilities.

Item 15. ***Recent Sales of Unregistered Securities.***

Set forth below is information regarding shares issued by us within the past three years. Also included is the consideration received by us for such shares and information relating to the section of the Securities Act, or rule of the SEC, under which exemption from registration was claimed.

Issuance of Securities.

In September 2015, we issued and sold 713,667 Class A common shares to our employees and a consultant at a price per share of $0.000273235 for aggregate gross consideration of $195.

In October 2015, we issued 3,568,353 Class B common shares to all of our Class A common shareholders as a distribution on their Class A common shares.

In October 2015, we issued and sold an aggregate of 8,028,809 Series A preferred shares to investors at a price per share of $4.6707 for aggregate gross consideration of $37.5 million.

In September 2016, we issued and sold 9,099,311 Series A preferred shares to investors at a price per share of $4.6707 for aggregate gross consideration of $42.5 million.

In March 2017, we issued and sold an aggregate of 5,757,372 Series B preferred shares to investors at a price per share of $6.9475 for aggregate gross consideration of $40.0 million.

In February 2018, we issued and sold an aggregate of 12,784,601 Series C preferred shares to investors at a price per $15.6438 per share for aggregate gross proceeds of $200.0 million.

Since July 15, 2015, the date of formation of the registrant, the registrant has issued 6,309 Class A common shares pursuant to the exercise of share options at an exercise price of $1.59 and 4,574 Class A common shares pursuant to the exercise of share options at an exercise price of $3.80, for total aggregate proceeds to the registrant of $27,375.

The securities listed above were issued pursuant to written compensatory plans or arrangements with our employees, directors and consultants, in reliance on the exemption provided by Rule 701 promulgated under the Securities Act, or pursuant to Section 4(a)(2) under the Securities Act, relative to transactions by an issuer not involving any public offering, to the extent an exemption from such registration was required.

Item 16. ***Exhibits and Financial Statement Schedules.***

(a) Exhibits.

Exhibit Number	Description of Exhibit
1.1	Form of Underwriting Agreement
3.1*	Memorandum of Association of the Registrant
3.2*	Amended and Restated Bye-laws of the Registrant (currently in effect)
3.3	Form of Amended and Restated Bye-laws of the Registrant (to be effective immediately following the closing of this offering)
4.1	Specimen Share Certificate evidencing the Class A common shares
4.2*	Second Amended and Restated Investors' Rights Agreement, dated as of February 9, 2018
5.1	Opinion of Conyers Dill & Pearman
10.1*	2015 Equity Incentive Plan, as amended, and form of share option grant notice and option agreement thereunder
10.2	2018 Incentive Award Plan and form of award agreements thereunder
10.3	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Sanj K. Patel, to be entered into in connection with this offering
10.4	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Stephen Mahoney, to be entered into in connection with this offering
10.5	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and John F. Paolini, to be entered into in connection with this offering
10.6†	Asset Purchase Agreement, dated September 7, 2016, by and between the Registrant and Biogen MA Inc., as amended
10.7†*	License Agreement, dated September 25, 2017, by and between the Registrant and Regeneron Pharmaceuticals, Inc.
10.8†	License Agreement, dated as of December 21, 2017, by and between the Registrant and MedImmune, Limited
10.9*	Clinical Supply Agreement, dated as of September 27, 2017, by and between the Registrant and Regeneron Pharmaceuticals, Inc.
10.10*	Sublease Agreement, dated as of March 13, 2018, by and between Kiniksa Pharmaceuticals Corp. and Shire Human Genetic Therapies, Inc.
10.11*	Form of Indemnification Agreement for Non-Fund-Designated Directors
10.12*	Form of Indemnification Agreement for Fund-Designated Directors
10.13*	Form of Indemnification Agreement for Officers
10.14	2018 Employee Share Purchase Plan

Exhibit Number	Description of Exhibit
10.15	Non-Employee Director Compensation Program
10.16	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Thomas Beetham, to be entered into in connection with this offering
10.17	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Chris Heberlig, to be entered into in connection with this offering
10.18	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Carsten Boess, to be entered into in connection with this offering
10.19	Form of Amended and Restated Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Rasmus Holm-Jorgensen, to be entered into in connection with this offering
21.1*	Subsidiaries of the Registrant
23.1	Consent of PricewaterhouseCoopers LLP, independent registered public accounting firm
23.2	Consent of Conyers Dill & Pearman (included in Exhibit 5.1)
24.1*	Power of Attorney

* Previously filed.

† Portions of this exhibit (indicated by asterisks) have been omitted pursuant to a request for confidential treatment pursuant to Rule 406 under the Securities Act of 1933, as amended.

(b) Financial Statement Schedules. Schedules not listed above have been omitted because the information required to be set forth therein is not applicable or is shown in the financial statements or notes thereto.

Item 17. ***Undertakings.***

The undersigned registrant hereby undertakes to provide to the underwriter, at the closing specified in the underwriting agreement, certificates in such denominations and registered in such names as required by the underwriter to permit prompt delivery to each purchaser.

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to directors, officers and controlling persons of the registrant pursuant to the foregoing provisions, or otherwise, the registrant has been advised that in the opinion of the SEC such indemnification is against public policy as expressed in the Securities Act and is, therefore, unenforceable. In the event that a claim for indemnification against such liabilities (other than the payment by the registrant of expenses incurred or paid by a director, officer or controlling person of the registrant in the successful defense of any action, suit or proceeding) is asserted by such director, officer or controlling person in connection with the securities being registered, the registrant will, unless in the opinion of its counsel the matter has been settled by controlling precedent, submit to a court of appropriate jurisdiction the question whether such indemnification by it is against public policy as expressed in the Securities Act and will be governed by the final adjudication of such issue.

The undersigned hereby undertakes that:

(1) For purposes of determining any liability under the Securities Act, the information omitted from the form of prospectus filed as part of this registration statement in reliance upon Rule 430A and contained in a form of prospectus filed by the registrant pursuant to Rule 424(b)(1) or (4) or 497(h) under the Securities Act shall be deemed to be part of this registration statement as of the time it was declared effective.

(2) For the purpose of determining any liability under the Securities Act, each post-effective amendment that contains a form of prospectus shall be deemed to be a new registration statement relating to the securities offered therein, and the offering of such securities at that time shall be deemed to be the initial *bona fide* offering thereof.

SIGNATURES

Pursuant to the requirements of the Securities Act of 1933, the registrant has duly caused this registration statement to be signed on its behalf by the undersigned, thereunto duly authorized in Hamilton, Bermuda, on this 14th day of May 2018.

KINIKSA PHARMACEUTICALS, LTD.

By: /s/ SANJ K. PATEL

Sanj K. Patel
Chief Executive Officer and Chairman of the Board of Directors

SIGNATURES

Pursuant to the requirements of the Securities Act of 1933, this registration statement has been signed by the following persons in the capacities and on the dates indicated.

Signature	Title	Date
/s/ SANJ K. PATEL Sanj K. Patel	Chief Executive Officer and Chairman of the Board of Directors (principal executive officer)	May 14, 2018
/s/ CHRIS HEBERLIG Chris Heberlig	Chief Financial Officer (principal financial and accounting officer)	May 14, 2018
* Felix J. Baker	Director	May 14, 2018
* Stephen R. Biggar	Director	May 14, 2018
* Thomas R. Malley	Director	May 14, 2018
* Tracey L. McCain	Director	May 14, 2018
* Kimberly J. Popovits	Director	May 14, 2018
* Barry D. Quart	Director	May 14, 2018

*By: /s/ SANJ K. PATEL
Attorney-in-fact

SIGNATURES

Pursuant to the requirements of the Securities Act of 1933, as amended, the undersigned, the duly authorized representative in the United States of the Registrant has signed this registration statement, on this 14th day of May 2018.

KINIKSA PHARMACEUTICALS, CORP.

By: /s/ SANJ K. PATEL

Sanj K. Patel
Chief Executive Officer

Exhibit 1.1

Kiniksa Pharmaceuticals, Ltd.

Class A Common Shares

Underwriting Agreement

, 2018

Goldman Sachs & Co. LLC,
J.P. Morgan Securities LLC
As representatives (the "Representatives") of the several Underwriters named in Schedule I hereto,

c/o Goldman Sachs & Co. LLC
200 West Street,
New York, NY 10282-2198.

c/o J.P. Morgan Securities LLC
383 Madison Avenue
New York, NY 10179

Ladies and Gentlemen:

Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company (the "Company"), proposes, subject to the terms and conditions stated in this agreement (this "Agreement"), to issue and sell to the Underwriters named in Schedule I hereto (the "Underwriters") an aggregate of [—] Class A common shares (the "Firm Shares") and, at the election of the Underwriters, up to [—] additional Class A common shares (the "Optional Shares") of the Company (such Class A common shares of the Company being referred to herein as the "Common Shares"). The Firm Shares and the Optional Shares that the Underwriters elect to purchase pursuant to Section 2 hereof are collectively called the "Shares".

1. The Company represents and warrants to, and agrees with, each of the Underwriters that:

(a) A registration statement on Form S-1 (File No. 333-[—]) (the "Initial Registration Statement") in respect of the Shares has been filed with the Securities and Exchange Commission (the "Commission"); the Initial Registration Statement and any post-effective amendment thereto, each in the form heretofore delivered to you, and excluding exhibits thereto, have been declared effective by the Commission in such form; other than a registration statement, if any, increasing the size of the offering (a "Rule 462(b) Registration Statement"), filed pursuant to Rule 462(b) under the Securities Act of 1933, as amended (the "Act"), which became effective upon filing, no other document with respect to the Initial Registration Statement has heretofore been filed with the Commission; and no stop order suspending the effectiveness of the Initial Registration

Statement, any post-effective amendment thereto or the Rule 462(b) Registration Statement, if any, has been issued and no proceeding for that purpose or pursuant to Section 8A of the Act has been initiated or threatened by the Commission (any preliminary prospectus included in the Initial Registration Statement or filed with the Commission pursuant to Rule 424(a) of the rules and regulations of the Commission under the Act is hereinafter called a "Preliminary Prospectus"; the various parts of the Initial Registration Statement and the Rule 462(b) Registration Statement, if any, including all exhibits thereto and including the information contained in the form of final prospectus filed with the Commission pursuant to Rule 424(b) under the Act in accordance with Section 5(a) hereof and deemed by virtue of Rule 430A under the Act to be part of the Initial Registration Statement at the time it was declared effective, each as amended at the time such part of the Initial Registration Statement became effective or such part of the Rule 462(b) Registration Statement, if any, became or hereafter becomes effective, are hereinafter collectively called the "Registration Statement"; the Preliminary Prospectus relating to the Shares that was included in the Registration Statement immediately prior to the Applicable Time (as defined in Section 1(c) hereof) is hereinafter called the "Pricing Prospectus"; such final prospectus, in the form first filed pursuant to Rule 424(b) under the Act, is hereinafter called the "Prospectus"; any oral or written communication with potential investors undertaken in reliance on Section 5(d) of the Act is hereinafter called a "Section 5(d) Communication"; any Section 5(d) Communication that is a written communication within the meaning of Rule 405 under the Act is hereinafter called a "Section 5(d) Writing"; and any "issuer free writing prospectus" as defined in Rule 433 under the Act relating to the Shares is hereinafter called an "Issuer Free Writing Prospectus");

(b) (A) No order preventing or suspending the use of any Preliminary Prospectus or any Issuer Free Writing Prospectus has been issued by the Commission, and (B) each Preliminary Prospectus, at the time of filing thereof, conformed in all material respects to the requirements of the Act and the rules and regulations of the Commission thereunder, and did not contain an untrue statement of a material fact or omit to state a material fact required to be stated therein or necessary to make the statements therein, in the light of the circumstances under which they were made, not misleading; *provided*, *however*, that this representation and warranty shall not apply to any statements or omissions made in reliance upon and in conformity with the Underwriter Information (as defined in Section 9(b) of this Agreement);

(c) For the purposes of this Agreement, the "Applicable Time" is [—] p.m. (Eastern time) on the date of this Agreement. The Pricing Prospectus, as supplemented by the information listed on Schedule II(c) hereto, taken together (collectively, the "Pricing Disclosure Package"), as of the Applicable Time, did not, and as of each Time of Delivery (as defined in Section 4(a) of this Agreement) will not, include any untrue statement of a material fact or omit to state any material fact necessary in order to make the statements therein, in the light of the circumstances under which they were made, not misleading; and each Issuer Free Writing Prospectus and each Section 5(d) Writing does not conflict with the information contained in the Registration Statement, the Pricing Prospectus or the Prospectus and each such Issuer Free Writing Prospectus and each Section 5(d) Writing, as supplemented by and taken together with the Pricing Disclosure Package, as of the Applicable Time, did not include any untrue statement of a material fact or omit to state any material fact necessary in order to make the statements therein, in the light of

the circumstances under which they were made, not misleading; provided, however, that this representation and warranty shall not apply to statements or omissions made in the Pricing Disclosure Package, any Issuer Free Writing Prospectus or any Section 5(d) Writing in reliance upon and in conformity with the Underwriter Information expressly for use therein;

(d) The Registration Statement conforms, and the Prospectus and any further amendments or supplements to the Registration Statement and the Prospectus will conform, in all material respects to the requirements of the Act and the rules and regulations of the Commission thereunder. The Registration Statement does not, and the Prospectus and any further amendments or supplements to the Registration Statement and the Prospectus will not, as of the applicable effective date as to each part of the Registration Statement and, as of the applicable filing date as to the Prospectus and any amendment or supplement thereto, contain an untrue statement of a material fact or omit to state a material fact required to be stated therein or necessary to make the statements therein not misleading; provided, however, that this representation and warranty shall not apply to any statements or omissions made in reliance upon and in conformity with the Underwriter Information expressly for use therein;

(e) Neither the Company nor any of its subsidiaries, when taken together as a whole, has sustained, since the date of the latest audited financial statements included in the Pricing Prospectus, (i) any material loss or material interference with its business from fire, explosion, flood or other calamity, whether or not covered by insurance, or from any labor dispute or court or governmental action, order or decree or (ii) entered into any transaction or agreement (whether or not in the ordinary course of business) that is material to the Company and its subsidiaries taken as a whole or incurred any liability or obligation, direct or contingent, that is material to the Company and its subsidiaries taken as a whole, in each case otherwise than as set forth or contemplated in the Pricing Prospectus; and, since the respective dates as of which information is given in the Registration Statement and the Pricing Prospectus, (i) there has not been any change in the share capital (other than as a result of (x) the exercise of any share options or the award of share options or restricted shares in the ordinary course of business pursuant to the Company's equity plans that are described in the Pricing Prospectus and the Prospectus or (y) the issuance, if any, of shares upon conversion of Company securities as described in the Pricing Prospectus and the Prospectus) or long-term debt of the Company or any of its subsidiaries (ii) or any Material Adverse Effect (as defined below); as used in this Agreement, "Material Adverse Effect" shall mean any material adverse change or effect, or any development involving a prospective material adverse change or effect, in or affecting (i) the business, properties, general affairs, management, financial position, shareholders' equity (deficit) or results of operations of the Company and its subsidiaries, taken together as a whole, except as set forth or contemplated in the Pricing Prospectus, or (ii) the ability of the Company to perform its obligations under this Agreement, including the issuance and sale of the Shares, or to consummate the transactions contemplated in the Pricing Prospectus and the Prospectus;

(f) Neither the Company nor any of its subsidiaries owns any real property. The Company and its subsidiaries have good and marketable title to all tangible personal property owned by them, in each case free and clear of all liens, encumbrances and defects except such as do not materially affect the value of such property and do not

interfere with the use made and proposed to be made of such property by the Company or its subsidiaries; and any real property and buildings held under lease by the Company or its subsidiaries are held, to the Company's knowledge, under valid, subsisting and enforceable leases with such exceptions as are not material and do not interfere with the use made and proposed to be made of such property and buildings by the Company or its subsidiaries;

(g) Each of the Company and each of its subsidiaries has been (i) duly organized and is validly existing and in good standing under the laws of its jurisdiction of organization, with power and authority (corporate and other) to own and/or lease its properties and conduct its business as described in the Pricing Prospectus, and (ii) duly qualified as a foreign corporation for the transaction of business and is in good standing under the laws of each other jurisdiction in which it owns or leases properties or conducts any business so as to require such qualification, except, in the case of this clause (ii), where the failure to have such power or authority or to be so qualified or in good standing would not, individually or in the aggregate, have a Material Adverse Effect, and each Significant Subsidiary (as defined in Rule 1-02(x) of Regulation S-X under the Act) of the Company has been listed in Exhibit 21.1 to the Registration Statement;

(h) The Company has not failed to make any filing with any Bermuda governmental authority, or to pay any Bermuda government fee or tax, which would make it liable to be struck off the Register of Companies and thereby cease to exist under the laws of Bermuda;

(i) The Company has an authorized capitalization as set forth in the Pricing Prospectus under the column labeled "Actual" under the caption "Capitalization" and all of the issued share capital of the Company has been duly authorized and validly issued and is fully paid and non-assessable and conforms to the description of the Common Shares contained in the Pricing Prospectus and Prospectus; and all of the issued share capital of each subsidiary of the Company has been duly and validly authorized and issued, is fully paid and non-assessable and is owned directly or indirectly by the Company, free and clear of all liens, encumbrances, equities or claims;

(j) The Shares to be issued and sold by the Company to the Underwriters hereunder have been duly authorized and, when issued and delivered against payment therefor as provided herein, will be duly authorized, validly issued, fully paid and non-assessable and will conform to the description of the Common Shares contained in the Pricing Disclosure Package and the Prospectus; and the issuance of the Shares is not subject to any preemptive or similar rights that have not been complied with or otherwise waived;

(k) The issue and sale of the Shares and the compliance by the Company with this Agreement and the consummation by the Company of the transactions contemplated in this Agreement and the Pricing Prospectus will not (A) conflict with or result in a breach or violation of any of the terms or provisions of, or constitute a default under any indenture, mortgage, deed of trust, loan agreement or other agreement or instrument to which the Company or any of its subsidiaries is a party or by which the Company or any of its subsidiaries is bound or to which any of the property or assets of the Company or any of its subsidiaries is subject, (B) result in any violation of the provisions of the certificate of incorporation, memorandum of association or bye-laws (or other applicable organizational document) of the Company or any of its subsidiaries, or (C) result in any

violation of any statute or any judgment, order, rule or regulation of any court or governmental agency or body having jurisdiction over the Company or any of its subsidiaries or any of their properties, except, in the case of (A) and (C), as would not, individually or in the aggregate, reasonably be expected to have a Material Adverse Effect; and no consent, approval, authorization, order, registration or qualification of or with any such court or governmental agency or body is required for the issue and sale of the Shares or the consummation by the Company of the transactions contemplated by this Agreement, except for the registration under the Act of the Shares, the approval by the Financial Industry Regulatory Authority ("FINRA") of the underwriting terms and arrangements, the approval for listing the Shares on an exchange of The Nasdaq Stock Market LLC and such consents, approvals, authorizations, registrations, orders or qualifications as may be required under state securities or Blue Sky laws in connection with the purchase and distribution of the Shares by the Underwriters;

(l) Neither the Company nor any of its subsidiaries is (i) in violation of its certificate of incorporation, memorandum of association or bye-laws (or other applicable organizational document), (ii) in violation of any law or statute or any judgment, order, rule or regulation of any court or governmental or regulatory authority, or (iii) in default in the performance or observance of any material obligation, agreement, covenant or condition contained in any indenture, mortgage, deed of trust, loan agreement, lease or other agreement or instrument to which it is a party or by which it or any of its properties may be bound, except, in the case of the foregoing clauses (ii) and (iii), for such defaults or violations as would not, individually or in the aggregate, reasonably be expected to have a Material Adverse Effect;

(m) The statements set forth in the Pricing Prospectus and the Prospectus under (i) the caption "Description of Share Capital", insofar as they purport to constitute a summary of the terms of the Common Shares, and (ii) the captions "Business—Intellectual Property," "Business—Government Regulation" and "Underwriting", insofar as they purport to describe the provisions of the laws and documents referred to therein, are accurate and complete in all material respects;

(n) The statements in the Pricing Prospectus and the Prospectus under the caption "Material Bermuda and U.S. Federal Income Tax Considerations—Material U.S. Federal Income Tax Considerations to U.S. Holders", insofar as such statements purport to constitute summaries of United States federal income tax law and regulations or legal conclusions with respect thereto, constitute accurate summaries of the matters described therein in all material respects;

(o) Other than as set forth in the Pricing Prospectus, there are no legal or governmental proceedings pending to which the Company or any of its subsidiaries is a party or of which any property of the Company or any of its subsidiaries is the subject which, if determined adversely to the Company or any of its subsidiaries, would individually or in the aggregate have a Material Adverse Effect; and, to the Company's knowledge, no such proceedings are threatened or contemplated by governmental authorities or threatened by others;

(p) The Company is not and, after giving effect to the offering and sale of the Shares and the application of the proceeds thereof as described in the Pricing Disclosure Package, will not be an "investment company", as such term is defined in the Investment Company Act of 1940, as amended (the "Investment Company Act");

(q) At the time of filing the Initial Registration Statement, the Company was not and, as of the date hereof, is not an "ineligible issuer," as defined under Rule 405 under the Act;

(r) PricewaterhouseCoopers LLP, who have certified certain financial statements of the Company and its subsidiaries, is an independent public accountant as required by the Act and the rules and regulations of the Commission thereunder;

(s) The Company maintains a system of internal control over financial reporting (as such term is defined in Rule 13a-15(f) under the Securities Exchange Act of 1934, as amended (the "Exchange Act")) that (i) complies with the requirements of the Exchange Act applicable to the Company, (ii) has been designed by the Company's principal executive officer and principal financial officer, or under their supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles and (iii) is sufficient to provide reasonable assurance that (A) transactions are executed in accordance with the Company's management's general or specific authorization, (B) transactions are recorded as necessary to permit preparation of financial statements in conformity with generally accepted accounting principles and to maintain accountability for assets, (C) access to assets is permitted only in accordance with the Company's management's general or specific authorization and (D) the recorded accountability for assets is compared with the existing assets at reasonable intervals and appropriate action is taken with respect to any differences; the Company's internal control over financial reporting is effective and the Company is not aware of any material weaknesses or significant deficiencies in its internal control over financial reporting;

(t) Since the date of the latest audited financial statements included in the Pricing Prospectus, no material weakness or significant deficiency has been identified in the internal control over financial reporting of the Company, and there has been no change in the Company's internal control over financial reporting that has materially and adversely affected, or is reasonably likely to materially and adversely affect, the Company's internal control over financial reporting;

(u) The Company maintains disclosure controls and procedures (as such term is defined in Rule 13a-15(e) under the Exchange Act) that are designed to comply with the requirements of the Exchange Act; such disclosure controls and procedures have been designed to ensure that material information relating to the Company and its subsidiaries is made known to the Company's principal executive officer and principal financial officer by others within those entities; and such disclosure controls and procedures are effective;

(v) The Company and its subsidiaries have filed all material tax returns required to be filed through the date hereof, or have duly requested extensions thereof, and have paid all material taxes shown as due thereon; all such tax returns are true and correct in all material respects; and no material deficiencies for taxes of the Company or any of its subsidiaries have been assessed or proposed adversely by a tax authority;

(w) There are no transfer taxes, stamp duties, capital duties, stamp duty reserve tax or similar fees or charges are payable by or on behalf of the Underwriters to

the Bermuda government or any political subdivision or taxing authority thereof in connection with the sale and delivery of the Shares to or for the respective accounts of the Underwriters or the sale and delivery by the Underwriters of the Shares to the initial purchasers thereof;

(x) The Company or its subsidiaries own, or have obtained valid and enforceable licenses for, the patents and patent applications, copyrights, trademarks, trademark registrations, service marks, service mark registrations, trade names, service names and know-how (including trade secrets and other unpatented and/or unpatentable proprietary or confidential information, systems or procedures) and all other technology and intellectual property rights, in each case, owned or purported to be owned or licensed or purported to be licensed to it, and except as described in the Registration Statement, the Pricing Disclosure Package and the Prospectus, to the knowledge of the Company, the Company and its subsidiaries own or have valid, binding and enforceable licenses or other rights under the patents and patent applications, copyrights, trademarks, trademark registrations, service marks, service mark registrations, trade names, service names and know-how (including trade secrets and other unpatented and/or unpatentable proprietary or confidential information, systems or procedures) and all other technology and intellectual property rights necessary for, or material to the conduct of their businesses as currently conducted or as proposed to be conducted in the manner described in the Registration Statement, Pricing Disclosure Package and the Prospectus (collectively, the "Company Intellectual Property"); to the knowledge of the Company, the Company owns or has valid, binding and enforceable licenses or other rights to practice such Company Intellectual Property; the intellectual property owned by the Company or any of its subsidiaries is free and clear of all material liens and encumbrances; to the knowledge of the Company, all patents, trademarks and copyrights owned or licensed by the Company or any of its subsidiaries are valid, enforceable and subsisting; the Company and its subsidiaries have complied with the terms of each agreement pursuant to which material intellectual property has been licensed to the Company or any subsidiary, and all such agreements are in full force and effect. Other than as disclosed in the Registration Statement, Pricing Disclosure Package and the Prospectus, (i) neither the Company nor any of its subsidiaries is obligated to pay a material royalty, grant a license or provide other material consideration to any third party in connection with the Company Intellectual Property (except for vendors providing services to the Company and its subsidiaries in the ordinary course of business), (ii) no action, suit, claim or other proceeding is pending, or, to the knowledge of the Company, is threatened, alleging that the Company or any of its subsidiaries is infringing, misappropriating, diluting or otherwise violating any rights of others with respect to any of the Company's or any of its subsidiaries' product candidates, processes or intellectual property, (iii) no action, suit, claim or other proceeding is pending, or, to the knowledge of the Company, is threatened, challenging the validity, enforceability, scope, registration, ownership or use of any of the patents or patent applications included in the Company Intellectual Property, or challenging the Company's or any of its subsidiaries' rights in or to any Company Intellectual Property, (iv) neither the Company nor any of its subsidiaries has received notice of any claim of infringement, misappropriation or conflict with any asserted rights of others with respect to any of the Company's or any of its subsidiaries' product candidates, technology, processes or Company Intellectual Property, (v) to the knowledge of the Company, the development, manufacture, sale, and any currently proposed use of any of the product

candidates or processes of the Company or any of its subsidiaries referred to in the Pricing Prospectus or the Prospectus do not currently, and will not upon commercialization, infringe any right or valid and issued patent claim of any third party, (vi) no third party has any ownership right in or to any Company Intellectual Property that is owned by the Company or any of its subsidiaries, and no third party has any ownership right in or to any Company Intellectual Property licensed to the Company or any of its subsidiaries, other than any licensor to the Company or such subsidiary (including, for clarity, any licensor to such licensor) of such Company Intellectual Property, (vii) the Company and its subsidiaries have taken reasonable measures to protect their confidential information and trade secrets and to maintain and safeguard the Company Intellectual Property, including the execution of appropriate nondisclosure and confidentiality agreements, (viii) to the knowledge of the Company, no employee, consultant or independent contractor of the Company or any of its subsidiaries is in or has ever been in violation in any material respect of any term of any employment contract, invention assignment agreement, non-competition agreement, non-solicitation agreement, nondisclosure agreement or any restrictive covenant to or with a former employer or independent contractor where the basis of such violation relates to such employee's employment or independent contractor's engagement with the Company or any of its subsidiaries or actions undertaken while employed or engaged with the Company of any of its subsidiaries, and (ix) to the knowledge of the Company, there is no infringement by third parties of any material Company Intellectual Property;

(y) All material patents and patent applications owned by or licensed to the Company or any of its subsidiaries or under which the Company or any of its subsidiaries has rights have been duly and properly filed, prosecuted and maintained; no person having a duty of candor to the U.S. Patent and Trademark Office ("USPTO") with respect to the prosecution of such patents has breached such duty; and the Company is not aware of any facts required to be disclosed to the USPTO that were not disclosed to the USPTO and which could form the basis of a finding of invalidity with respect to any patents that have issued;

(z) The Company and each of its subsidiaries (i) is, and since each of their respective date of incorporation, has been in compliance with all statutes, rules, regulations, and policies applicable to the ownership, testing, development, registration, licensure, manufacture, processing, use, recordkeeping, filing of reports, storage, import, export or disposal of any product developed or manufactured by or on behalf of the Company or such subsidiary, including, without limitation, the U.S. Federal Food, Drug and Cosmetic Act (21 U.S.C. § 301 et seq.), the Public Health Service Act (42 U.S.C. § 201 et seq.), the U.S. Anti-Kickback Statute (42 U.S.C. § 1320a-7b(b)), the Health Insurance Portability and Accountability Act of 1996, as amended by the Health Information Technology for Economic and Clinical Health Act of 2009, the regulations promulgated pursuant to such laws, including, without limitation, 21 C.F.R. Parts 50, 54, 56, 58 and 312, the U.S. Animal Welfare Act and comparable state laws, and all other local, state, federal, national and foreign laws, rules, and regulations, applicable to the Company or any of its subsidiaries (collectively, the "Applicable Laws"), except where such non-compliance with Applicable Laws would not, individually or in the aggregate, have a Material Adverse Effect; (ii) has not received any written notice from any court or arbitrator or governmental or regulatory authority or third party alleging or asserting

material non-compliance with any Applicable Laws or any licenses, exemptions, certificates, approvals, clearances, authorizations, permits, registrations and supplements or amendments thereto required by any such Applicable Laws ("Authorizations"); (iii) possesses all material Authorizations and such Authorizations are valid and in full force and effect and are not in violation of any term of any such Authorizations, except where a failure to so possess, invalidity, a failure to be in full force and effect, or such violation would not, individually or in the aggregate, have a Material Adverse Effect; (iv) has not received written notice of any claim, action, suit, proceeding, hearing, enforcement, investigation, arbitration or other action, or notice of adverse finding, any FDA Form 483, warning letter, untitled letter or other communication, from any governmental or regulatory authority, in each case, alleging or asserting material noncompliance with any Applicable Laws or Authorizations nor, to the knowledge of the Company, is any such claim, action, suit, proceeding, hearing, enforcement, investigation, arbitration or other action or communication threatened; (v) has not since each of their respective date of incorporation received written notice from any court or arbitrator or governmental or regulatory authority that such court, arbitrator or authority has taken, is taking or intends to take action to materially limit, suspend, materially modify or revoke any Authorizations nor, to the knowledge of the Company, is any such limitation, suspension, modification or revocation threatened, except where such limitation, suspension, modification or revocation would not, individually or in the aggregate, have a Material Adverse Effect; (vi) has filed, obtained, maintained or submitted all material reports, documents, forms, notices, applications, records, claims, submissions and supplements or amendments as required by any Applicable Laws or Authorizations and that all such reports, documents, forms, notices, applications, records, claims, submissions and supplements or amendments were complete and accurate on the date filed in all material respects (or were corrected or supplemented by a subsequent submission), except where such failure would not, individually or in the aggregate, have a Material Adverse Effect; and (vii) is not a party to any corporate integrity agreements, monitoring agreements, consent decrees, settlement orders, or similar agreements with or imposed by any governmental or regulatory authority;

(aa) (i) The clinical trials and pre-clinical studies conducted by or on behalf of or sponsored by the Company or any of its subsidiaries or in which the Company or any of its subsidiaries has participated, and that are either described in the Pricing Prospectus and the Prospectus, or the results of which are referred to in the Pricing Prospectus or the Prospectus, or that are intended to be submitted to the European Medicines Agency (the "EMA"), the U.S. Food and Drug Administration ("FDA") or similar foreign, state, and local governmental or regulatory authorities (collectively, the "Regulatory Authorities") as a basis for product approval, were and, if still pending, are being conducted in all material respects in accordance with all Applicable Laws; (ii) the descriptions in the Pricing Prospectus and the Prospectus of the results of such trials and studies are accurate and complete in all material respects and fairly present the data derived from such trials and studies; (iii) the Company has no knowledge of any other trials the results of which are materially inconsistent with or otherwise could reasonably be expected to materially discredit or call into question the results described or referred to in the Pricing Prospectus or the Prospectus; and (iv) neither the Company nor any of its subsidiaries has received any written notices, correspondence or other written communication from any Regulatory Authority or any other governmental agency or any institutional review board or other

similar entity which could lead to the termination, suspension or material modification of any clinical or pre-clinical trials that are described in the Pricing Prospectus or the Prospectus or the results of which are referred to in the Pricing Prospectus or the Prospectus;

(bb) The Company and each of its subsidiaries has filed for and received all material Authorizations issued by, and has made all declarations and filings with, the applicable Regulatory Authorities that are necessary for the ownership or lease of its properties or the conduct of its business as currently conducted as described in the Pricing Prospectus and the Prospectus, or to permit the clinical trials as currently conducted by or on behalf of the Company or any of its subsidiaries, including, without limitation, all necessary EMA, FDA and other applicable regulatory agency Authorizations outside of the United States as applicable to the Company's current clinical activities; and no event has occurred which allows, or after notice or lapse of time would allow, revocation, termination or modification of any such Authorization or result in any other material impairment of the rights of the holder of any such Authorization;

(cc) Neither the Company nor any of its subsidiaries, nor any of its or any of its subsidiaries' officers, directors or managing employees (as defined in 42 U.S.C. § 1320a-5(b)), is or has been excluded, suspended or debarred by FDA or comparable authorities pursuant to 21 U.S.C. 335a or similar laws, or from participation in any national or regional health care program, or made subject to any pending or, to the knowledge of the Company, threatened or contemplated action which could reasonably be expected to result in such exclusion, suspension or debarment;

(dd) This Agreement has been duly authorized, executed and delivered by the Company;

(ee) None of the Company or any of its subsidiaries or its or their directors or officers nor, to the knowledge of the Company, any agent, employee, affiliate or other person associated with or acting on behalf of the Company or any of its subsidiaries has (i) made, offered, promised or authorized any unlawful contribution, gift, entertainment or other unlawful expense; (ii) made, offered, promised or authorized any direct or indirect unlawful payment; or (iii) violated or is in violation of any provision of the Foreign Corrupt Practices Act of 1977, the Bribery Act 2010 of the United Kingdom or any other applicable anti-bribery or anti-corruption law. The Company and its subsidiaries have instituted, maintain and enforce, and will continue to maintain and enforce policies and procedures designed to promote and ensure compliance with all applicable anti-bribery and anti-corruption laws;

(ff) The operations of the Company and its subsidiaries are and have been conducted at all times in compliance with the requirements of applicable anti-money laundering laws, including, but not limited to, the Bank Secrecy Act of 1970, as amended by the USA PATRIOT ACT of 2001, and the rules and regulations promulgated thereunder, and the anti-money laundering laws of the various jurisdictions in which the Company and its subsidiaries conduct business (collectively, the "Money Laundering Laws") and no action, suit or proceeding by or before any court or governmental agency, authority or body or any arbitrator involving the Company or any of its subsidiaries with

respect to the Money Laundering Laws is pending or, to the knowledge of the Company, threatened;

(gg) None of the Company or any of its subsidiaries or its or their directors or officers nor, to the knowledge of the Company, any agent, employee or affiliate of the Company or any of its subsidiaries is currently the subject or the target of any sanctions administered or enforced by the U.S. Government, including, without limitation, the Office of Foreign Assets Control of the U.S. Department of the Treasury ("OFAC"), or the U.S. Department of State and including, without limitation, the designation as a "specially designated national" or "blocked person," the European Union, Her Majesty's Treasury, the United Nations Security Council, or other relevant sanctions authority (collectively, "Sanctions"), and the Company will not directly or indirectly use the proceeds of the offering of the Shares hereunder, or lend, contribute or otherwise make available such proceeds to any subsidiary, joint venture partner or other person or entity (i) to fund or facilitate any activities of or business with any person, or in any country or territory, that, at the time of such funding, is the subject or the target of Sanctions or (ii) in any other manner that will result in a violation by any person (including any person participating in the transaction, whether as underwriter, advisor, investor or otherwise) of Sanctions. For the past five years, the Company has not knowingly engaged in and are not now knowingly engaged in any dealings or transactions with any person that at the time of the dealing or transaction is or was the subject or the target of Sanctions or with any country or territory that is the subject or target of Sanctions;

(hh) The Company and its directors and officers have taken all necessary actions to ensure that, upon the effectiveness of the Registration Statement, the Company will be in compliance with all provisions of the Sarbanes-Oxley Act of 2002 and the rules and regulations promulgated thereunder that the Company is required to comply with as of the effectiveness of the Registration Statement;

(ii) (1) The legality, validity, enforceability or admissibility of evidence of this Agreement and (2) the admissibility into evidence of the Registration Statement, the Pricing Disclosure Package and the Prospectus in any jurisdiction in which the Company is organized or does business is, in either case, not dependent upon such document being submitted into, filed or recorded with any court or other authority in any such jurisdiction on or before the date hereof or that any tax, imposition or charge be paid in any such jurisdiction on or in respect of any such document;

(jj) The indemnification and contribution provisions set forth in Section 9 hereof do not contravene Bermuda law or public policy;

(kk) Subject to the qualifications, limitations, exceptions and assumptions set forth in the Preliminary Prospectus and the Prospectus, the Company does not believe that it was a passive foreign investment company (a "PFIC"), as defined in section 1297 of the Internal Revenue Code of 1986, as amended;

(ll) Any final judgment for a fixed or determined sum of money rendered by any U.S. federal or New York state court located in the State of New York having jurisdiction under its own laws in respect of any suit, action or proceeding against the Company based upon this Agreement, the Registration Statement, the Pricing Package and the Prospectus would be declared enforceable against the Company by the courts of Bermuda, without reconsideration or reexamination of the merits;

(mm) Neither the Company nor any of its subsidiaries or their properties or assets has immunity under Bermuda, U.S. federal or New York state law from any legal action, suit or proceeding, from the giving of any relief in any such legal action, suit or proceeding, from set-off or counterclaim, from the jurisdiction of any Bermuda, U.S. federal or New York state court, from service of process, attachment upon or prior to judgment, or attachment in aid of execution of judgment, or from execution of a judgment, or other legal process or proceeding for the giving of any relief or for the enforcement of a judgment, in any such court with respect to their respective obligations, liabilities or any other matter under or arising out of or in connection herewith; and, to the extent that the Company or any of its subsidiaries or any of its properties, assets or revenues may have or may hereafter become entitled to any such right of immunity in any such court in which proceedings arising out of, or relating to the transactions contemplated by this Agreement, the Registration Statement, the Pricing Package and the Prospectus, may at any time be commenced, the Company has waived, and it will waive, or will cause its subsidiaries to waive, such right to the extent permitted by law;

(nn) The choice of laws of the State of New York as the governing law of this Agreement, the Registration Statement, the Pricing Package and the Prospectus is a valid choice of law under the laws of Bermuda and will be recognized and given effect to in any such action brought before a court of competent jurisdiction in Bermuda, except for those laws (i) which such court considers to be procedural in nature, (ii) which are revenue or penal laws, or (iii) the application of which would be inconsistent with public policy, as such term is interpreted under the laws of Bermuda. The Company has the power to submit, and pursuant to Section 18 of this Agreement, has legally, validly, effectively and irrevocably submitted, to the personal jurisdiction of each New York State and United States federal court sitting in the Borough of Manhattan in the City of New York and has validly and irrevocably waived any objection to the laying of venue of any suit, action or proceeding brought in such court;

(oo) A holder of the Shares and each Underwriter has standing to bring an action before the appropriate courts in Bermuda for the enforcement of their respective rights under this Agreement and the Shares. It is not necessary or advisable in order for any of the Underwriters or a holder of Shares to enforce their respective rights under this Agreement, including the exercise of the remedies thereunder, that it be licensed, qualified or otherwise entitled to carry on business in Bermuda;

(pp) Any statistical and market-related data included in the Pricing Prospectus or the Prospectus are based on or derived from sources that the Company believes, after reasonable inquiry, to be reliable and accurate and, to the extent required, the Company has obtained the written consent to the use of such data from such sources;

(qq) Neither the Company nor any of its subsidiaries has taken and the Company will not take, and will cause its subsidiaries not to take, directly or indirectly, without giving effect to activities by the Underwriters, any action that is designed to or that has constituted or which would reasonably be expected to cause or result in stabilization or manipulation of the price of the Shares;

(rr) Except as described in the Pricing Prospectus, there are no contracts, agreements or understandings between the Company or any of its subsidiaries and any person granting such person the right to require the Company or any subsidiary to file a

registration statement under the Act with respect to any securities of the Company or any subsidiary owned or to be owned by such person;

(ss) Except pursuant to this Agreement, there are no contracts, agreements or understandings between the Company or any of its subsidiaries and any person that would give rise to a valid claim against the Company, any of its subsidiaries or any Underwriter for a brokerage commission, finder's fee or other like payment in connection with the offering of the Shares;

(tt) There are no debt securities or preferred shares of, or guaranteed by, the Company that are rated by a "nationally recognized statistical rating organization," as such term is defined by Section 3(a)(62) of the Exchange Act;

(uu) The Company and each of its subsidiaries are insured by insurers of recognized financial responsibility against such losses and risks and in such amounts as are, in the Company's reasonable judgment, prudent and customary in the businesses in which they are engaged; and neither the Company nor any of its subsidiaries has any reason to believe that it will not be able to renew its existing insurance coverage as and when such coverage expires or to obtain similar coverage from similar insurers as may be necessary to continue its business;

(vv) Except as disclosed in the Pricing Prospectus, there are no off-balance sheet arrangements (as defined in Regulation S-K Item 303(a)(4)(ii)) that may have a material current or future effect on the Company's financial condition, changes in financial condition, results of operations, liquidity, capital expenditures or capital resources;

(ww) No subsidiary of the Company is currently prohibited, directly or indirectly, under any agreement or other instrument to which it is a party or is subject, from paying any dividends to the Company, from making any other distribution on such subsidiary's share capital, from repaying to the Company any loans or advances to such subsidiary from the Company or from transferring any or all of such subsidiary's properties or assets to the Company or any other subsidiary of the Company. No governmental or regulatory approvals, consents, authorizations orders, licenses, registrations, clearances or qualifications are currently required in Bermuda or from any political subdivisions of the Bermuda government in order for the Company to pay dividends or other distributions declared by the Company to the holders of the Shares. All dividends and other distributions declared and payable on the Shares may under the current laws and regulations of Bermuda be freely transferred out of Bermuda, subject to any applicable Money Laundering Laws or Sanctions. No such dividends or other distributions are subject to withholding or other taxes under the current laws and regulations of Bermuda, in each case except as described in the Registration Statement, the Pricing Package and the Prospectus;

(xx) The financial statements included in the Registration Statement, the Pricing Disclosure Package and the Prospectus, together with the related schedules and notes, present fairly in all material respects the financial position of the Company and its subsidiaries as of the dates indicated, and for the periods indicated therein, subject in the case of unaudited financial statements, to normal year-end audit adjustments; said financial statements have been prepared in conformity with U.S. generally accepted accounting principles ("GAAP") applied on a consistent basis throughout the periods involved. The supporting schedules, if any, present fairly in all material respects in

accordance with GAAP the information required to be stated therein. The selected financial data and the summary financial information included in the Registration Statement, the Pricing Disclosure Package and the Prospectus present fairly in all material respects the information shown therein and have been compiled on a basis consistent with that of the audited financial statements included therein. Except as included therein, no historical or pro forma financial statements or supporting schedules are required to be included in the Registration Statement, the Pricing Disclosure Package or the Prospectus under the Act or the rules and regulations promulgated thereunder; and

(yy) From the time of initial confidential submission of a registration statement relating to the Shares with the Commission (or, if earlier, the first date on which a Section 5(d) Communication was made) through the date hereof, the Company has been and is an "emerging growth company" as defined in Section 2(a)(19) of the Act (an "Emerging Growth Company").

2. Subject to the terms and conditions herein set forth, the Company agrees to issue and sell to each of the Underwriters, and each of the Underwriters agrees, severally and not jointly, to purchase from the Company, at a purchase price per share of $[—], the number of Firm Shares set forth opposite the name of such Underwriter in Schedule I hereto and (b) in the event and to the extent that the Underwriters shall exercise the election to purchase Optional Shares as provided below, the Company agrees to issue and sell to each of the Underwriters, and each of the Underwriters agrees, severally and not jointly, to purchase from the Company, at the purchase price per share set forth in clause (a) of this Section 2 (provided that the purchase price per Optional Share shall be reduced by an amount per share equal to any dividends or distributions declared by the Company and payable on the Firm Shares but not payable on the Optional Shares), that portion of the number of Optional Shares as to which such election shall have been exercised (to be adjusted by you so as to eliminate fractional shares) determined by multiplying such number of Optional Shares by a fraction, the numerator of which is the maximum number of Optional Shares which such Underwriter is entitled to purchase as set forth opposite the name of such Underwriter in Schedule I hereto and the denominator of which is the maximum number of Optional Shares that all of the Underwriters are entitled to purchase hereunder.

The Company hereby grants to the Underwriters the right to purchase at their election up to [—] Optional Shares, at the purchase price per share set forth in the paragraph above, for the sole purpose of covering sales of shares in excess of the number of Firm Shares, provided that the purchase price per Optional Share shall be reduced by an amount per share equal to any dividends or distributions declared by the Company and payable on the Firm Shares but not payable on the Optional Shares. Any such election to purchase Optional Shares may be exercised only by written notice from you to the Company, given within a period of 30 calendar days after the date of this Agreement, setting forth the aggregate number of Optional Shares to be purchased and the date on which such Optional Shares are to be delivered, as determined by you but in no event earlier than the First Time of Delivery (as defined in Section 4 hereof) or, unless you and the Company otherwise agree in writing, earlier than two or later than ten business days after the date of such notice.

3. Upon the authorization by you of the release of the Firm Shares, the several Underwriters propose to offer the Firm Shares for sale upon the terms and conditions set forth in the Pricing Prospectus and the Prospectus.

4. (a) The Shares to be purchased by each Underwriter hereunder, in electronic form, and in such authorized denominations and registered in such names as the Representatives may request upon at least forty-eight hours' prior notice to the Company shall be delivered by or on behalf of the Company to the Representatives, through the facilities of the Depository Trust Company ("DTC"), for the account of such Underwriter, against payment by or on behalf of such Underwriter of the purchase price therefor by wire transfer of Federal (same-day) funds to the account specified by the Company to the Representatives at least forty-eight hours in advance. . The time and date of such delivery and payment shall be, with respect to the Firm Shares, 9:30 a.m., New York City time, on [—], 2018 or such other time and date as the Representatives and the Company may agree upon in writing, and, with respect to the Optional Shares, 9:30 a.m., New York City time, on the date specified by the Representatives in each written notice given by the Representatives of the Underwriters' election to purchase such Optional Shares, or such other time and date as the Representatives and the Company may agree upon in writing. Such time and date for delivery of the Firm Shares is herein called the "First Time of Delivery", such time and date for delivery of the Optional Shares, if not the First Time of Delivery, is herein called the "Second Time of Delivery", and each such time and date for delivery is herein called a "Time of Delivery".

(b) The documents to be delivered at each Time of Delivery by or on behalf of the parties hereto pursuant to Section 8 hereof, including the cross receipt for the Shares and any additional documents requested by the Underwriters pursuant to Section 8(n) hereof, will be delivered at the offices of Ropes & Gray LLP, Prudential Tower, 800 Boylston Street, Boston, Massachusetts 02199 (the "Closing Location"), and the Shares will be delivered at the Office of DTC or its designated location, all at such Time of Delivery. A meeting will be held at the Closing Location at [—]p.m., New York City time, on the New York Business Day next preceding such Time of Delivery, at which meeting the final drafts of the documents to be delivered pursuant to the preceding sentence will be available for review by the parties hereto. For the purposes of this Section 4, "New York Business Day" shall mean each Monday, Tuesday, Wednesday, Thursday and Friday which is not a day on which banking institutions in New York City are generally authorized or obligated by law or executive order to close.

5. The Company agrees with each of the Underwriters:

(a) To prepare the Prospectus in a form approved by you and to file such Prospectus pursuant to Rule 424(b) under the Act not later than the Commission's close of business on the second business day following the execution and delivery of this Agreement, or, if applicable, such earlier time as may be required by Rule 430A(a)(3) under the Act; to make no further amendment or any supplement to the Registration Statement or the Prospectus prior to the last Time of Delivery which shall be disapproved by you promptly after reasonable notice thereof; to advise you, promptly after it receives notice thereof, of the time when any amendment to the Registration Statement has been filed or becomes effective or any amendment or supplement to the Prospectus has been filed and to furnish you with copies thereof; to file promptly all material required to be filed by the Company with the Commission pursuant to Rule 433(d) under the Act; to advise

you, promptly after it receives notice thereof, of the issuance by the Commission of any stop order or of any order preventing or suspending the use of any Registration Statement, Preliminary Prospectus or other prospectus in respect of the Shares, of the suspension of the qualification of the Shares for offering or sale in any jurisdiction, of the initiation or threatening of any proceeding for any such purpose or pursuant to Section 8A of the Act, or of any request by the Commission for the amending or supplementing of the Registration Statement or the Prospectus or for additional information; and, in the event of the issuance of any stop order or of any order preventing or suspending the use of any Preliminary Prospectus or other prospectus relating to the Shares or suspending any such qualification, to promptly use its best efforts to obtain the withdrawal of such order;

(b) Promptly from time to time to take such action as you may reasonably request to qualify the Shares for offering and sale under the securities laws of such jurisdictions as you may request and to comply with such laws so as to permit the continuance of sales and dealings therein in such jurisdictions for as long as may be necessary to complete the distribution of the Shares, provided that in connection therewith the Company shall not be required to qualify as a foreign corporation or to file a general consent to service of process in any jurisdiction, or subject itself to taxation in any such jurisdiction in which it was not otherwise subject to taxation;

(c) Prior to 10:00 a.m., New York City time, on the New York Business Day next succeeding the date of this Agreement (or such other time as may be agreed to by the Representatives and the Company) and from time to time, to furnish the Underwriters with written and electronic copies of the Prospectus, Preliminary Prospectus and any supplements and amendments thereto or to the Registration Statement in such quantities as the Representatives may reasonably request, and, if the delivery of a prospectus (or in lieu thereof, the notice referred to in Rule 173(a) under the Act) is required at any time prior to the expiration of nine months after the time of issue of the Prospectus in connection with the offering or sale of the Shares and if at such time any event shall have occurred as a result of which the Prospectus as then amended or supplemented would include an untrue statement of a material fact or omit to state any material fact necessary in order to make the statements therein, in the light of the circumstances under which they were made when such Prospectus (or in lieu thereof, the notice referred to in Rule 173(a) under the Act) is delivered, not misleading, or, if for any other reason it shall be necessary during such same period to amend or supplement the Prospectus in order to comply with the Act, to notify you and, before amending or supplementing the Registration Statement, the Pricing Disclosure Package or the Prospectus, to furnish you a copy of each such proposed amendment or supplement and not file any such proposed amendment or supplement to which you reasonably object, and upon your request to prepare and furnish without charge to each Underwriter and to any dealer in securities (whose name and address the Underwriters shall furnish to the Company) as many written and electronic copies as you may from time to time reasonably request of an amended Prospectus or a supplement to the Prospectus which will correct such statement or omission or effect such compliance; and in case any Underwriter is required to deliver a prospectus (or in lieu thereof, the notice referred to in Rule 173(a) under the Act) in connection with sales of any of the Shares at any time nine months or more after the time of issue of the Prospectus, upon your request but at the expense of such Underwriter, to prepare and deliver to such Underwriter as many written and electronic

copies as you may request of an amended or supplemented Prospectus complying with Section 10(a)(3) of the Act;

(d) To make generally available to its securityholders as soon as practicable (which may be satisfied by filing with the Commission's Electronic Data Gathering Analysis and Retrieval System ("EDGAR")), but in any event not later than sixteen months after the effective date of the Registration Statement (as defined in Rule 158(c) under the Act), an earnings statement of the Company (which need not be audited) complying with Section 11(a) of the Act and the rules and regulations of the Commission thereunder (including, at the option of the Company, Rule 158);

(e)(1) During the period beginning from the date hereof and continuing to and including the date 180 days after the date of the Prospectus (the "Lock-Up Period"), not to (i) offer, sell, contract to sell, pledge, grant any option to subscribe or purchase, make any short sale or otherwise transfer or dispose of, directly or indirectly, or file with or confidentially submit to the Commission a registration statement under the Act relating to, any securities of the Company that are substantially similar to the Shares, including but not limited to any options or warrants to purchase Common Shares or any securities that are convertible into or exchangeable for, or that represent the right to receive, Common Shares or any such substantially similar securities, or publicly disclose the intention to make any offer, sale, pledge, disposition, submission or filing or (ii) enter into any swap or other agreement that transfers, in whole or in part, any of the economic consequences of ownership of the Common Shares or any such other securities, whether any such transaction described in clause (i) or (ii) above is to be settled by delivery of Common Shares or such other securities, in cash or otherwise, without the prior written consent of the Representatives. The foregoing restrictions shall not apply to (A) the Shares to be sold hereunder, (B) grants of awards pursuant to equity incentive plans existing as of the date of this Agreement and disclosed in the Pricing Prospectus, (C) issuances of Common Shares pursuant to the exercise of awards granted pursuant to clause (B), (D) the filing of any registration statement on Form S-8 relating to any benefit plans or arrangements disclosed in the Pricing Prospectus and the issuance of securities registered pursuant thereto, (E) issuances of securities under the Company's employee share purchase plan existing as of the date of this Agreement and described in the Pricing Prospectus, (F) issuances of Common Shares or securities exercisable for, convertible into or exchangeable for Common Shares in connection with any acquisition, collaboration, licensing or other joint venture or strategic transaction or any debt financing transaction involving the Company and an unaffiliated third party and that includes a bona fide commercial relationship, and (G) the issuance of securities pursuant to an agreement described in the Pricing Prospectus; provided that, in the case of clause (F) such issuances shall not in the aggregate be greater than 5% of the total outstanding Common Shares of the Company outstanding immediately following the completion of this offering of Shares; and provided further that, in the case of clauses (B), (C), (E) and (F) the Company shall (i) cause each recipient of such securities to execute and deliver to you, on or prior to the issuance of such securities, a lock-up agreement on substantially the same terms as the lock-up agreements referenced in Section 8(k) hereof for the remainder of the Company Lock-Up Period, and (ii) enter stop transfer instructions with the Company's transfer agent and registrar on such securities, which the Company agrees it will not waive or amend without the prior written consent of each Representative.

(e)(2) If the Representatives, in their sole discretion, agree to release or waive the restrictions set forth in a lock-up letter described in Section 8(k) hereof for an officer or director of the Company and provide the Company with notice of the impending release or waiver at least three business days before the effective date of the release or waiver, the Company agrees to announce the impending release or waiver by a press release substantially in the form of Annex I hereto through a major news service at least two business days before the effective date of the release or waiver;

(f) During a period of three years from the effective date of the Registration Statement, so long as the Company is subject to the reporting requirements of either Section 13 or Section 15(d) of the Exchange Act, to furnish to its shareholders as soon as practicable after the end of each fiscal year an annual report (including a balance sheet and statements of income, shareholders' equity and cash flows of the Company and its consolidated subsidiaries certified by independent public accountants) and, as soon as practicable after the end of each of the first three quarters of each fiscal year (beginning with the fiscal quarter ending after the effective date of the Registration Statement), to make available to its shareholders consolidated summary financial information of the Company and its subsidiaries for such quarter in reasonable detail; *provided* that any report, communication or financial statement furnished or filed with the Commission that is publicly available on the Commission's EDGAR system shall be deemed to have been furnished to the Company's shareholders at the time furnished or filed with the Commission;

(g) During a period of three years from the effective date of the Registration Statement, to furnish to you copies of all reports or other communications (financial or other) furnished to shareholders, and to deliver to you (i) as soon as they are available, copies of any reports and financial statements furnished to or filed with the Commission or any national securities exchange on which any class of securities of the Company is listed and (ii) such additional information concerning the business and financial condition of the Company as you may from time to time reasonably request (such financial statements to be on a consolidated basis to the extent the accounts of the Company and its subsidiaries are consolidated in reports furnished to its shareholders generally; *provided* that any report, communication or financial statement furnished or filed with the Commission that is publicly available on the Commission's EDGAR system shall be deemed to have been furnished to you at the time furnished or filed with the Commission;

(h) To use the net proceeds received by it from the sale of the Shares pursuant to this Agreement in the manner specified in the Pricing Prospectus under the caption "Use of Proceeds";

(i) To use its best efforts to list for trading, subject to official notice of issuance, the Shares on the Nasdaq Stock Market ("Nasdaq");

(j) To file with the Commission such information on Form 10-Q or Form 10-K as may be required by Rule 463 under the Act;

(k) If the Company elects to rely upon Rule 462(b), the Company shall file a Rule 462(b) Registration Statement with the Commission in compliance with Rule 462(b) by 10:00 P.M., Washington, D.C. time, on the date of this Agreement, and the Company shall at the time of filing either pay to the Commission the filing fee for the Rule 462(b)

Registration Statement or give irrevocable instructions for the payment of such fee pursuant to Rule 111(b) under the Act;

(l) Upon request of any Underwriter, to furnish, or cause to be furnished, to such Underwriter an electronic version of the Company's trademarks, servicemarks and corporate logo for use on the website, if any, operated by such Underwriter for the purpose of facilitating the on-line offering of the Shares (the "License"); provided, however, that the License shall be used solely for the purpose described above, is granted without any fee and may not be assigned or transferred; and

(m) To promptly notify you if the Company ceases to be an Emerging Growth Company at any time prior to the later of (i) completion of the distribution of the Shares within the meaning of the Act and (ii) completion of the Lock-Up Period referred to in Section 5(e) hereof.

6. (a) The Company represents and agrees that, without the prior consent of the Representatives, it has not made and will not make any offer relating to the Shares that would constitute a "free writing prospectus" as defined in Rule 405 under the Act; each Underwriter represents and agrees that, without the prior consent of the Company and the Representatives, it has not made and will not make any offer relating to the Shares that would constitute a free writing prospectus required to be filed with the Commission; any such free writing prospectus the use of which has been consented to by the Company and the Representatives is listed on Schedule II(a) hereto;

(b) The Company has complied and will comply with the requirements of Rule 433 under the Act applicable to any Issuer Free Writing Prospectus, including timely filing with the Commission or retention where required and legending; and the Company represents that it has satisfied and agrees that it will satisfy the conditions under Rule 433 under the Act to avoid a requirement to file with the Commission any electronic road show;

(c) The Company agrees that if at any time following issuance of an Issuer Free Writing Prospectus or Section 5(d) Writing any event occurred or occurs as a result of which such Issuer Free Writing Prospectus or Section 5(d) Writing would conflict with the information in the Registration Statement, the Pricing Prospectus or the Prospectus or would include an untrue statement of a material fact or omit to state any material fact necessary in order to make the statements therein, in the light of the circumstances then prevailing, not misleading, the Company will give prompt notice thereof to the Representatives and, if requested by the Representatives, will prepare and furnish without charge to each Underwriter an Issuer Free Writing Prospectus, Section 5(d) Writing or other document which will correct such conflict, statement or omission; provided, however, that this covenant shall not apply to any statements or omissions in an Issuer Free Writing Prospectus or Section 5(d) Writing made in reliance upon and in conformity with the Underwriter Information;

(d) The Company represents and agrees that (i) it has not engaged in, or authorized any other person to engage in, any Section 5(d) Communications, other than Section 5(d) Communications with the prior consent of the Representatives with entities that are qualified institutional buyers as defined in Rule 144A under the Act or institutions that are accredited investors as defined in Rule 501(a) under the Act; and (ii) it has not

distributed, or authorized any other person to distribute, any Section 5(d) Writings, other than those distributed with the prior consent of the Representatives that are listed on Schedule II(b) hereto; and the Company reconfirms that the Underwriters have been authorized to act on its behalf in engaging in Section 5(d) Communications; and

(e) Each Underwriter represents and agrees that any Section 5(d) Communications undertaken by it were with entities that are qualified institutional buyers as defined in Rule 144A under the Act or institutions that are accredited investors as defined in Rule 501(a) under the Act.

7. The Company covenants and agrees with the several Underwriters that the Company will pay or cause to be paid the following: (i) the fees, disbursements and expenses of any of the Company's counsel and accountants in connection with the registration of the Shares under the Act and all other expenses in connection with the preparation, printing, reproduction and filing of the Registration Statement, any Preliminary Prospectus, any Section 5(d) Writing, any Issuer Free Writing Prospectus and the Prospectus and amendments and supplements thereto and the mailing and delivering of copies thereof to the Underwriters and dealers; (ii) the cost of printing or producing any Agreement among Underwriters, this Agreement, the Blue Sky Memorandum, closing documents (including any compilations thereof) and any other documents in connection with the offering, purchase, sale and delivery of the Shares; (iii) all expenses in connection with the qualification of the Shares for offering and sale under state or foreign securities laws as provided in Section 5(b) hereof, including the fees and disbursements of counsel for the Underwriters in connection with such qualification and in connection with the Blue Sky survey (iv) all fees and expenses in connection with listing the Shares on Nasdaq; (v) the filing fees incident to, and the fees and disbursements of counsel for the Underwriters in connection with, any required review by FINRA of the terms of the sale of the Shares; (vi) the cost of preparing share certificates; (vii) the cost and charges of any transfer agent or registrar; and (viii) all other costs and expenses of the Company related to investor presentations on any "road show" undertaken in connection with the marketing of the Shares, including without limitation, expenses associated with the production of road show slides and graphics, fees and expenses of any consultants engaged in connection with the road show presentations, travel and lodging expenses of the representatives and officers of the Company, any such consultants and the Underwriters and their representatives (provided, however, that the Underwriters and the Company shall each pay 50% of the cost of chartering any aircraft to be used in connection with the road show by the Company and the Underwriters) and (ix) all other costs and expenses incident to the performance of its obligations hereunder which are not otherwise specifically provided for in this Section; provided, however, that the amount payable by the Company pursuant to subsections (iii) and (v) of this Section 7, for the fees and disbursements of counsel to the Underwriters described in such subsections, shall not exceed $30,000 in the aggregate. It is understood, however, that, except as provided in this Section, and Sections 9 and 12 hereof, the Underwriters will pay all of their own costs and expenses, including the fees of their counsel, share transfer taxes on resale of any of the Shares by them, and any advertising expenses connected with any offers they may make.

8. The obligations of the Underwriters hereunder, as to the Shares to be delivered at each Time of Delivery, shall be subject, in their discretion, to the condition that all representations and warranties and other statements of the Company herein are, at and as of the date hereof and such Time of Delivery, true and correct, the condition that the Company shall have performed all of its obligations hereunder theretofore to be performed, and the following additional conditions:

(a) The Prospectus shall have been filed with the Commission pursuant to Rule 424(b) under the Act within the applicable time period prescribed for such filing by the rules and regulations under the Act and in accordance with Section 5(a) hereof; all material required to be filed by the Company pursuant to Rule 433(d) under the Act shall have been filed with the Commission within the applicable time period prescribed for such filing by Rule 433; if the Company has elected to rely upon Rule 462(b) under the Act, the Rule 462(b) Registration Statement shall have become effective by 10:00 P.M., Washington, D.C. time, on the date of this Agreement; no stop order suspending the effectiveness of the Registration Statement or any part thereof shall have been issued and no proceeding for that purpose shall have been initiated or threatened by the Commission; no stop order suspending or preventing the use of the Pricing Prospectus, Prospectus or any Issuer Free Writing Prospectus shall have been initiated or threatened by the Commission; and all requests for additional information on the part of the Commission shall have been complied with to your reasonable satisfaction;

(b) Ropes & Gray LLP, counsel for the Underwriters, shall have furnished to you their written opinion and negative assurance letter, dated such Time of Delivery, in form and substance satisfactory to you, and such counsel shall have received such papers and information as they may reasonably request to enable them to pass upon such matters;

(c) Taylors in association with Walkers, Bermuda counsel for the Underwriters, shall have furnished to you their written opinion, dated such Time of Delivery, in form and substance satisfactory to you;

(d) Latham & Watkins LLP, counsel for the Company, shall have furnished to you their written opinion and negative assurance letter, dated such Time of Delivery, in form and substance satisfactory to you;

(e) Latham & Watkins LLP, special tax counsel for the Company, shall have furnished to you their written opinion, dated such Time of Delivery, in form and substance satisfactory to you;

(f) Conyers Dill & Pearman Limited, Bermuda counsel for the Company, shall have furnished to you their written opinion, dated such Time of Delivery, in form and substance satisfactory to you;

(g) Proskauer Rose LLP, intellectual property counsel for the Company, shall have furnished to you their written opinion and negative assurance letter, each dated such Time of Delivery, in form and substance satisfactory to you;

(h) On the date of the Prospectus at a time prior to the execution of this Agreement, at 9:30 a.m., New York City time, on the effective date of any

post-effective amendment to the Registration Statement filed subsequent to the date of this Agreement and also at each Time of Delivery, PricewaterhouseCoopers LLP shall have furnished to you a letter or letters, dated the respective dates of delivery thereof, in form and substance satisfactory to you, containing statements and information of the type ordinarily included in accountants' "comfort letters" to underwriters with respect to the financial statements and certain financial information contained in the Registration Statement, the Pricing Prospectus and the Prospectus; provided that the letter delivered on the Time of Delivery shall use a "cut-off date" not earlier than the date of the Prospectus;

(i) (i) Neither the Company nor any of its subsidiaries shall have sustained since the date of the latest audited financial statements included in the Pricing Prospectus any loss or interference with its business from fire, explosion, flood or other calamity, whether or not covered by insurance, or from any labor dispute or court or governmental action, order or decree, otherwise than as set forth or contemplated in the Pricing Prospectus, and (ii) since the respective dates as of which information is given in the Pricing Prospectus there shall not have been any change in the share capital or long-term debt of the Company or any of its subsidiaries or any change or effect, or any development involving a prospective change or effect, in or affecting (x) the business, properties, general affairs, management, financial position, shareholders' equity or results of operations of the Company and its subsidiaries, taken as a whole, except as set forth or contemplated in the Pricing Prospectus and the Prospectus, or (y) the ability of the Company to perform its obligations under this Agreement, including the issuance and sale of the Shares, or to consummate the transactions contemplated in the Pricing Prospectus and the Prospectus, the effect of which, in any such case described in clause (i) or (ii), is in your judgment so material and adverse as to make it impracticable or inadvisable to proceed with the public offering or the delivery of the Shares being delivered at such Time of Delivery on the terms and in the manner contemplated in the Pricing Prospectus and the Prospectus;

(j) On or after the Applicable Time there shall not have occurred any of the following: (i) a suspension or material limitation in trading in securities generally on the New York Stock Exchange or Nasdaq; (ii) a suspension or material limitation in trading in the Company's securities on Nasdaq; (iii) a general moratorium on commercial banking activities declared by either Federal, New York or Massachusetts State or Bermuda authorities or a material disruption in commercial banking or securities settlement or clearance services in the United States; (iv) the outbreak or escalation of hostilities involving the United States or the declaration by the United States of a national emergency or war or (v) the occurrence of any other calamity or crisis or any change in financial, political or economic conditions in the United States or elsewhere, if the effect of any such event specified in clause (iv) or (v) in your judgment makes it impracticable or inadvisable to proceed with the public offering or the delivery of the Shares being delivered at such Time of Delivery on the terms and in the manner contemplated in the Pricing Prospectus;

(k) The Shares to be sold at such Time of Delivery shall have been duly listed on Nasdaq;

(l) The Company shall have obtained and delivered to the Underwriters executed copies of an agreement from the directors, officers, employees and equityholders of the Company, substantially to the effect set forth in Section 5(e) hereof in form and substance satisfactory to you;

(m) The Company shall have complied with the provisions of Section 5(c) hereof with respect to the furnishing of prospectuses;

(n) The Company shall have furnished or caused to be furnished to you at such Time of Delivery a certificate of the chief financial officer of the Company, in form and substance reasonably satisfactory to you;

(o) The Company shall have furnished or caused to be furnished to you at such Time of Delivery certificates of officers of the Company satisfactory to you as to the accuracy of the representations and warranties of the Company herein at and as of such Time of Delivery, as to the performance by the Company of all of its obligations hereunder to be performed at or prior to such Time of Delivery, as to the matters set forth in subsections (a) and (e) of this Section and as to such other matters as you may reasonably request; and

(p) The Company shall have furnished or caused to be furnished to you, at such Time of Delivery, such additional information, certificates, opinions or documents as the Representatives may reasonably request.

9. (a) The Company will indemnify and hold harmless each Underwriter against any losses, claims, damages or liabilities, joint or several, to which such Underwriter may become subject, under the Act or otherwise, insofar as such losses, claims, damages or liabilities (or actions in respect thereof) arise out of or are based upon an untrue statement or alleged untrue statement of a material fact contained in the Registration Statement, any Preliminary Prospectus, the Pricing Prospectus or the Prospectus, or any amendment or supplement thereto, any Issuer Free Writing Prospectus, any "roadshow" as defined in Rule 433(h) under the Act (a "roadshow"), or any "issuer information" filed or required to be filed pursuant to Rule 433(d) under the Act or any Section 5(d) Writing, or arise out of or are based upon the omission or alleged omission to state therein a material fact required to be stated therein or necessary to make the statements therein not misleading, and will reimburse each Underwriter for any legal or other expenses reasonably incurred by such Underwriter in connection with investigating or defending any such action or claim as such expenses are incurred; *provided*, *however*, that the Company shall not be liable in any such case to the extent that any such loss, claim, damage or liability arises out of or is based upon an untrue statement or alleged untrue statement or omission or alleged omission made in the Registration Statement, any Preliminary Prospectus, the Pricing Prospectus or the Prospectus, or any amendment or supplement thereto, or any Issuer Free Writing Prospectus or any Section 5(d) Writing, in reliance upon and in conformity with the Underwriter Information expressly for use therein.

(b) Each Underwriter, severally and not jointly, will indemnify and hold harmless the Company against any losses, claims, damages or liabilities to which the Company may become subject, under the Act or otherwise, insofar as such losses, claims,

damages or liabilities (or actions in respect thereof) arise out of or are based upon an untrue statement or alleged untrue statement of a material fact contained in the Registration Statement, any Preliminary Prospectus, the Pricing Prospectus or the Prospectus, or any amendment or supplement thereto, or any Issuer Free Writing Prospectus, or any roadshow or Section 5(d) Writing, or arise out of or are based upon the omission or alleged omission to state therein a material fact required to be stated therein or necessary to make the statements therein not misleading, in each case to the extent, but only to the extent, that such untrue statement or alleged untrue statement or omission or alleged omission was made in the Registration Statement, any Preliminary Prospectus, the Pricing Prospectus or the Prospectus, or any amendment or supplement thereto, or any Issuer Free Writing Prospectus, or any roadshow or Section 5(d) Writing, in reliance upon and in conformity with the Underwriter Information; and will reimburse the Company for any legal or other expenses reasonably incurred by the Company in connection with investigating or defending any such action or claim as such expenses are incurred. As used in this Agreement with respect to an Underwriter and an applicable document, "Underwriter Information" shall mean the written information furnished to the Company by such Underwriter through the Representatives expressly for use therein; it being understood and agreed upon that the only such information furnished by any Underwriter consists of the following information in the Prospectus furnished on behalf of each Underwriter: the concession and reallowance figures appearing in the [—] paragraph under the caption "Underwriting", and the information contained in the [—] paragraph under the caption "Underwriting".

(c) Promptly after receipt by an indemnified party under subsection (a) or (b) above of notice of the commencement of any action, such indemnified party shall, if a claim in respect thereof is to be made against the indemnifying party under such subsection, notify the indemnifying party in writing of the commencement thereof; provided that the failure to notify the indemnifying party shall not relieve it from any liability that it may have under the preceding paragraphs of this Section 9 except to the extent that it has been materially prejudiced (through the forfeiture of substantive rights or defenses) by such failure; and provided further that the failure to notify the indemnifying party shall not relieve it from any liability that it may have to an indemnified party otherwise than under the preceding paragraphs of this Section 9. In case any such action shall be brought against any indemnified party and it shall notify the indemnifying party of the commencement thereof, the indemnifying party shall be entitled to participate therein and, to the extent that it shall wish, jointly with any other indemnifying party similarly notified, to assume the defense thereof, with counsel satisfactory to such indemnified party (who shall not, except with the consent of the indemnified party, be counsel to the indemnifying party), and, after notice from the indemnifying party to such indemnified party of its election so to assume the defense thereof, the indemnifying party shall not be liable to such indemnified party under such subsection for any legal expenses of other counsel, in each case subsequently incurred by such indemnified party, in connection with the defense thereof other than reasonable costs of investigation. In any such proceeding, any indemnified party shall have the right to retain its own counsel, but the fees and expenses of such counsel shall be at the expense of such indemnified party unless (i) the indemnifying party and the indemnified party shall have mutually agreed to the contrary; (ii) the indemnifying party has failed within a reasonable time to retain counsel reasonably satisfactory to the indemnified party; (iii) the indemnified

party shall have reasonably concluded that there may be legal defenses available to it that are different from or in addition to those available to the indemnifying party; or (iv) the named parties in any such proceeding (including any impleaded parties) include both the indemnifying party and the indemnified party and representation of both parties by the same counsel would be inappropriate due to actual or potential differing interests between them. It is understood and agreed that the indemnifying party shall not, in connection with any proceeding or related proceeding in the same jurisdiction, be liable for the fees and expenses of more than one separate firm (in addition to any local counsel) for all indemnified parties, and that all such fees and expenses shall be paid or reimbursed as they are incurred. No indemnifying party shall, without the written consent of the indemnified party, effect the settlement or compromise of, or consent to the entry of any judgment with respect to, any pending or threatened action or claim in respect of which indemnification or contribution may be sought hereunder (whether or not the indemnified party is an actual or potential party to such action or claim) unless such settlement, compromise or judgment (i) includes an unconditional release of the indemnified party from all liability arising out of such action or claim and (ii) does not include a statement as to or an admission of fault, culpability or a failure to act, by or on behalf of any indemnified party.

(d) If the indemnification provided for in this Section 9 is unavailable to or insufficient to hold harmless an indemnified party under subsection (a) or (b) above in respect of any losses, claims, damages or liabilities (or actions in respect thereof) referred to therein, then each indemnifying party shall contribute to the amount paid or payable by such indemnified party as a result of such losses, claims, damages or liabilities (or actions in respect thereof) in such proportion as is appropriate to reflect the relative benefits received by the Company on the one hand and the Underwriters on the other from the offering of the Shares. If, however, the allocation provided by the immediately preceding sentence is not permitted by applicable law, then each indemnifying party shall contribute to such amount paid or payable by such indemnified party in such proportion as is appropriate to reflect not only such relative benefits but also the relative fault of the Company on the one hand and the Underwriters on the other in connection with the statements or omissions which resulted in such losses, claims, damages or liabilities (or actions in respect thereof), as well as any other relevant equitable considerations. The relative benefits received by the Company on the one hand and the Underwriters on the other shall be deemed to be in the same proportion as the total net proceeds from the offering (before deducting

expenses) received by the Company bear to the total underwriting discounts and commissions received by the Underwriters, in each case as set forth in the table on the cover page of the Prospectus. The relative fault shall be determined by reference to, among other things, whether the untrue or alleged untrue statement of a material fact or the omission or alleged omission to state a material fact relates to information supplied by the Company on the one hand or the Underwriters on the other and the parties' relative intent, knowledge, access to information and opportunity to correct or prevent such statement or omission. The Company and the Underwriters agree that it would not be just and equitable if contribution pursuant to this subsection (d) were determined by *pro rata* allocation (even if the Underwriters were treated as one entity for such purpose) or by any other method of allocation which does not take account of the equitable considerations referred to above in this subsection (d). The amount paid or payable by an indemnified party as a result of

the losses, claims, damages or liabilities (or actions in respect thereof) referred to above in this subsection (d) shall be deemed to include any legal or other expenses reasonably incurred by such indemnified party in connection with investigating or defending any such action or claim. Notwithstanding the provisions of this subsection (d), no Underwriter shall be required to contribute any amount in excess of the amount by which the total price at which the Shares underwritten by it and distributed to the public were offered to the public exceeds the amount of any damages which such Underwriter has otherwise been required to pay by reason of such untrue or alleged untrue statement or omission or alleged omission. No person guilty of fraudulent misrepresentation (within the meaning of Section 11(f) of the Act) shall be entitled to contribution from any person who was not guilty of such fraudulent misrepresentation. The Underwriters' obligations in this subsection (d) to contribute are several in proportion to their respective underwriting obligations and not joint.

(e) The obligations of the Company under this Section 9 shall be in addition to any liability which the Company may otherwise have and shall extend, upon the same terms and conditions, to each officer and director of each Underwriter and each person, if any, who controls any Underwriter within the meaning of the Act and each broker-dealer affiliate of any Underwriter; and the obligations of the Underwriters under this Section 9 shall be in addition to any liability which the respective Underwriters may otherwise have and shall extend, upon the same terms and conditions, to each officer and director of the Company (including any person who, with his or her consent, is named in the Registration Statement as about to become a director of the Company) and to each person, if any, who controls the Company within the meaning of the Act.

10. (a) If any Underwriter shall default in its obligation to purchase the Shares which it has agreed to purchase hereunder at a Time of Delivery, you may in your discretion arrange for you or another party or other parties to purchase such Shares on the terms contained herein. If within thirty-six hours after such default by any Underwriter you do not arrange for the purchase of such Shares, then the Company shall be entitled to a further period of thirty-six hours within which to procure another party or other parties satisfactory to you to purchase such Shares on such terms. In the event that, within the respective prescribed periods, you notify the Company that you have so arranged for the purchase of such Shares, or the Company notifies you that it has so arranged for the purchase of such Shares, you or the Company shall have the right to postpone such Time of Delivery for a period of not more than seven days, in order to effect whatever changes may thereby be made necessary in the Registration Statement or the Prospectus, or in any other documents or arrangements, and the Company agrees to file promptly any amendments or supplements to the Registration Statement or the Prospectus which in your opinion may thereby be made necessary. The term "Underwriter" as used in this Agreement shall include any person substituted under this Section with like effect as if such person had originally been a party to this Agreement with respect to such Shares.

(b) If, after giving effect to any arrangements for the purchase of the Shares of a defaulting Underwriter or Underwriters by you and the Company as provided in subsection (a) above, the aggregate number of such Shares which remains unpurchased does not exceed one-eleventh of the aggregate number of all the Shares to be purchased at such Time of Delivery, then the Company shall have the right to require each

non-defaulting Underwriter to purchase the number of Shares which such Underwriter agreed to purchase hereunder at such Time of Delivery and, in addition, to require each non-defaulting Underwriter to purchase its pro rata share (based on the number of Shares which such Underwriter agreed to purchase hereunder) of the Shares of such defaulting Underwriter or Underwriters for which such arrangements have not been made; but nothing herein shall relieve a defaulting Underwriter from liability for its default.

(c) If, after giving effect to any arrangements for the purchase of the Shares of a defaulting Underwriter or Underwriters by you and the Company as provided in subsection (a) above, the aggregate number of such Shares which remains unpurchased exceeds one-eleventh of the aggregate number of all the Shares to be purchased at such Time of Delivery, or if the Company shall not exercise the right described in subsection (b) above to require non-defaulting Underwriters to purchase Shares of a defaulting Underwriter or Underwriters, then this Agreement (or, with respect to the Second Time of Delivery, the obligations of the Underwriters to purchase and of the Company to sell the Optional Shares) shall thereupon terminate, without liability on the part of any non-defaulting Underwriter or the Company, except for the expenses to be borne by the Company and the Underwriters as provided in Section 7 hereof and the indemnity and contribution agreements in Section 9 hereof; but nothing herein shall relieve a defaulting Underwriter from liability for its default.

11. The respective indemnities, agreements, representations, warranties and other statements of the Company and the several Underwriters, as set forth in this Agreement or made by or on behalf of them, respectively, pursuant to this Agreement, shall remain in full force and effect, regardless of any investigation (or any statement as to the results thereof) made by or on behalf of any Underwriter or its affiliates or any officer, director or controlling person of any Underwriter, or the Company, or any officer or director or controlling person of the Company, and shall survive delivery of and payment for the Shares.

12. If this Agreement shall be terminated pursuant to Section 10 hereof, the Company shall not then be under any liability to any Underwriter except as provided in Sections 7 and 9 hereof; but, if for any other reason, any Shares are not delivered by or on behalf of the Company as provided herein, the Company will reimburse the Underwriters through you for all out-of-pocket expenses approved in writing by you, including reasonable fees and disbursements of counsel, reasonably incurred by the Underwriters in making preparations for the purchase, sale and delivery of the Shares not so delivered.

13. In all dealings hereunder, you shall act on behalf of each of the Underwriters, and the parties hereto shall be entitled to act and rely upon any statement, request, notice or agreement on behalf of any Underwriter made or given by you jointly or by Goldman Sachs & Co. LLC or J.P. Morgan Securities LLC on behalf of you as the representatives.

All statements, requests, notices and agreements hereunder shall be in writing, and if to the Underwriters shall be delivered or sent by mail, telex or facsimile transmission to each of you as the Representatives in care of Goldman Sachs & Co. LLC, 200 West Street, New York, New York 10282-2198, Attention: Registration Department; and J.P. Morgan Securities LLC, 383 Madison Avenue, New York, New York 10179,

Attention: [—]; and if to the Company shall be delivered or sent by mail, telex or facsimile transmission to the address of the Company set forth in the Registration Statement, Attention: Secretary; provided, however, that any notice to an Underwriter pursuant to Section 9(c) hereof shall be delivered or sent by mail, telex or facsimile transmission to such Underwriter at its address set forth in its Underwriters' Questionnaire, or telex constituting such Questionnaire, which address will be supplied to the Company by you upon request; provided, however, that notices under subsection 5(e) shall be in writing, and if to the Underwriters shall be delivered or sent by mail, telex or facsimile transmission to you as the Representatives at Goldman Sachs & Co. LLC, 200 West Street, New York, New York 10282-2198, Attention: Control Room. Any such statements, requests, notices or agreements shall take effect upon receipt thereof.

In accordance with the requirements of the USA Patriot Act (Title III of Pub. L. 107-56 (signed into law October 26, 2001)), the Underwriters are required to obtain, verify and record information that identifies their respective clients, including the Company, which information may include the name and address of their respective clients, as well as other information that will allow the Underwriters to properly identify their respective clients.

14. This Agreement shall be binding upon, and inure solely to the benefit of, the Underwriters, the Company and, to the extent provided in Sections 9 and 11 hereof, the officers and directors of the Company and each person who controls the Company or any Underwriter and the directors, officers and affiliates of the Underwriters, and their respective heirs, executors, administrators, successors and assigns, and no other person shall acquire or have any right under or by virtue of this Agreement. No purchaser of any of the Shares from any Underwriter shall be deemed a successor or assign by reason merely of such purchase.

15. Time shall be of the essence of this Agreement. As used herein, the term "business day" shall mean any day when the Commission's office in Washington, D.C. is open for business.

16. The Company acknowledges and agrees that (i) the purchase and sale of the Shares pursuant to this Agreement is an arm's-length commercial transaction between the Company, on the one hand, and the several Underwriters, on the other, (ii) in connection therewith and with the process leading to such transaction each Underwriter is acting solely as a principal and not the agent or fiduciary of the Company, (iii) no Underwriter has assumed an advisory or fiduciary responsibility in favor of the Company with respect to the offering contemplated hereby or the process leading thereto (irrespective of whether such Underwriter has advised or is currently advising the Company on other matters) or any other obligation to the Company except the obligations expressly set forth in this Agreement and (iv) the Company has consulted its own legal and financial advisors to the extent it deemed appropriate. The Company agrees that it will not claim that the Underwriters, or any of them, has rendered advisory services of any nature or respect, or owes a fiduciary or similar duty to the Company, in connection with such transaction or the process leading thereto.

17. This Agreement supersedes all prior agreements and understandings (whether written or oral) between the Company and the Underwriters, or any of them, with respect to the subject matter hereof.

18. THIS AGREEMENT AND ANY MATTERS ARISING OUT OF OR RELATED TO THIS TRANSACTION SHALL BE GOVERNED BY AND CONSTRUED IN ACCORDANCE WITH THE LAWS OF THE STATE OF NEW YORK WITHOUT REGARD TO PRINCIPLES OF CONFLICT OF LAWS THAT WOULD RESULT IN THE APPLICATION OF ANY LAW OTHER THAN THE LAWS OF THE STATE OF NEW YORK. The Company agrees that any suit or proceeding arising in respect of this Agreement or any transaction contemplated by this Agreement will be tried exclusively in the U.S. District Court for the Southern District of New York or, if that court does not have subject matter jurisdiction, in any state court located in The City and County of New York and the Company agrees to submit to the jurisdiction of, and to venue in, such courts.

19. The Company and each of the Underwriters hereby irrevocably waives, to the fullest extent permitted by applicable law, any and all right to trial by jury in any legal proceeding arising out of or relating to this Agreement or the transactions contemplated hereby.

20. This Agreement may be executed by any one or more of the parties hereto in any number of counterparts, each of which shall be deemed to be an original, but all such counterparts shall together constitute one and the same instrument.

21. The Company agrees to indemnify each Underwriter, its directors, officers, affiliates and each person, if any, who controls such Underwriter within the meaning of Section 15 of the Securities Act or Section 20 of the Exchange Act, against any loss incurred by such Underwriter as a result of any judgment or order being given or made for any amount due hereunder and such judgment or order being expressed and paid in a currency (the "judgment currency") other than U.S. dollars and as a result of any variation as between (i) the rate of exchange at which the U.S. dollar amount is converted into the judgment currency for the purpose of such judgment or order, and (ii) the rate of exchange at which such indemnified person is able to purchase U.S. dollars with the amount of the judgment currency actually received by the indemnified person. The foregoing indemnity shall constitute a separate and independent obligation of the Company and shall continue in full force and effect notwithstanding any such judgment or order as aforesaid. The term "rate of exchange" shall include any premiums and costs of exchange payable in connection with the purchase of, or conversion into, the relevant currency.

22. To the extent that the Company has or hereafter may acquire any immunity (sovereign or otherwise) from jurisdiction of any court of (i) Bermuda, or any political subdivision thereof, (ii) the United States or the State of New York, (iii) any jurisdiction in which it owns or leases property or assets or from any legal process (whether through service of notice, attachment prior to judgment, attachment in aid of execution, execution, set-off or otherwise) with respect to themselves or their respective property and assets or this Agreement, the Company hereby irrevocably waives such immunity in respect of its obligations under this Agreement to the fullest extent permitted by applicable law.

23. The Company irrevocably appoints Kiniksa Pharmaceuticals Corp., located in Lexington, Massachusetts, as its authorized agent in the Borough of Manhattan in The City of New York upon which process may be served in any such suit or proceeding, and agrees that service of process upon such authorized agent, and written notice of such service to the Company by the person serving the same to the address provided in this Section 23, shall be deemed in every respect effective service of process upon the

Company in any such suit or proceeding. The Company hereby represents and warrants that such authorized agent has accepted such appointment and has agreed to act as such authorized agent for service of process. The Company further agrees to take any and all action as may be necessary to maintain such designation and appointment of such authorized agent in full force and effect for a period of seven years from the date of this Agreement.

If the foregoing is in accordance with your understanding, please sign and return to us counterparts hereof, and upon the acceptance hereof by you, on behalf of each of the Underwriters, this letter and such acceptance hereof shall constitute a binding agreement between each of the Underwriters and the Company. It is understood that your acceptance of this letter on behalf of each of the Underwriters is pursuant to the authority set forth in a form of Agreement among Underwriters, the form of which shall be submitted to the

Company for examination upon request, but without warranty on your part as to the authority of the signers thereof.

Very truly yours,

Kiniksa Pharmaceuticals, Ltd.

By: _____
Name:
Title:

Accepted as of the date hereof:

Goldman Sachs & Co. LLC

By: _____
Name:
Title:

J.P. Morgan Securities LLC

By: _____
Name:
Title:

SCHEDULE I

Underwriter	Total Number of Firm Shares to be Purchased	Number of Optional Shares to be Purchased if Maximum Option Exercised
Goldman Sachs & Co. LLC		
J.P. Morgan Securities LLC		
JMP Securities LLC		
Wedbush Securities Inc.		
Total		

SCHEDULE II

(a) Issuer Free Writing Prospectuses not included in the Pricing Disclosure Package:
[—]

(b) Section 5(d) Writings:
[—]

(c) Information other than the Pricing Prospectus that comprise the Pricing Disclosure Package:

The initial public offering price per share for the Shares is $[—]
The number of Shares purchased by the Underwriters is [—].
[—]

ANNEX I

[Form of Press Release]

[Company]
[Date]

("[Company]") announced today that Goldman Sachs & Co. LLC and J.P. Morgan Securities LLC, the lead book-running managers in the Company's recent public sale of shares of common shares, are [waiving] [releasing] a lock-up restriction with respect to shares of the Company's common shares held by [certain officers or directors] [an officer or director] of the Company. The [waiver] [release] will take effect on , 20 , and the shares may be sold on or after such date.

This press release is not an offer for sale of the securities in the United States or in any other jurisdiction where such offer is prohibited, and such securities may not be offered or sold in the United States absent registration or an exemption from registration under the United States Securities Act of 1933, as amended.

Exhibit 3.3

AMENDED & RESTATED BYE-LAWS

OF

KINIKSA PHARMACEUTICALS, LTD.

TABLE OF CONTENTS

Kiniksa Pharamceuticals, Ltd.

INTERPRETATION

1. Definitions

1.1 In these Bye-laws, the following words and expressions shall, where not inconsistent with the context, have the following meanings, respectively:

Act	the Companies Act 1981;
Auditor	includes an individual, company or partnership;
Board	the board of directors (including, for the avoidance of doubt, a sole director) appointed or elected pursuant to these Bye-laws and acting by resolution in accordance with the Act and these Bye-laws or the directors present at a meeting of directors at which there is a quorum;
Company	the company for which these Bye-laws are approved and confirmed;
Director	a director of the Company;
Member	the person registered in the Register of Members as the holder of shares in the Company and, when two or more persons are so registered as joint holders of shares, means the person whose name stands first in the Register of Members as one of such joint holders or all of such persons, as the context so requires;
notice	written notice as further provided in these Bye-laws unless otherwise specifically stated;
Officer	any person appointed by the Board to hold an office in the Company;
Register of Directors and Officers	the register of directors and officers referred to in these Bye-laws;
Register of Members	the register of members referred to in these Bye-laws;
Resident Representative	any person appointed to act as resident representative and includes any deputy or assistant

	resident representative;
Secretary	the person appointed to perform any or all of the duties of secretary of the Company and includes any deputy or assistant secretary and any person appointed by the Board to perform any of the duties of the Secretary; and
Treasury Share	a share of the Company that was or is treated as having been acquired and held by the Company and has been held continuously by the Company since it was so acquired and has not been cancelled.

1.2 In these Bye-laws, where not inconsistent with the context:

(a) words denoting the plural number include the singular number and *vice versa*;

(b) words denoting the masculine gender include the feminine and neuter genders;

(c) words importing persons include companies, associations or bodies of persons whether corporate or not;

(d) the words:-

(i) "may" shall be construed as permissive; and

(ii) "shall" shall be construed as imperative;

(e) a reference to statutory provision shall be deemed to include any amendment or re-enactment thereof;

(f) the phrase "issued and outstanding" in relation to shares, means shares in issue other than Treasury Shares;

(g) the word "corporation" means a corporation whether or not a company within the meaning of the Act; and

(h) unless otherwise provided herein, words or expressions defined in the Act shall bear the same meaning in these Bye-laws.

1.3 In these Bye-laws expressions referring to writing or its cognates shall, unless the contrary intention appears, include facsimile, printing, lithography, photography, electronic mail and other modes of representing words in visible form.

1.4 Headings used in these Bye-laws are for convenience only and are not to be used or relied upon in the construction hereof.

SHARES

2. Power to Issue Shares

2.1 Subject to these Bye-laws and to any resolution of the Members to the contrary, and without prejudice to any special rights previously conferred on the holders of any existing shares or class of shares, the Board shall have the power to issue any unissued shares on such terms and conditions as it may determine.

2.2 Subject to the Act, any preference shares may be issued or converted into shares that (at a determinable date or at the option of the Company or the holder) are liable to be redeemed on such terms and in such manner as may be determined by the Board (before the issue or conversion).

3. Power of the Company to Purchase its Shares

3.1 The Company may purchase its own shares for cancellation or acquire them as Treasury Shares in accordance with the Act on such terms as the Board shall think fit.

3.2 The Board may exercise all the powers of the Company to purchase or acquire all or any part of its own shares in accordance with the Act.

4. Rights Attaching to Shares

Subject to any resolution of the Members to the contrary (and without prejudice to any special rights conferred thereby on the holders of any other shares or class of shares), the share capital shall be divided as follows:

4.1 The Company is authorised to issue five classes of shares to be designated "Class A Common Shares," "Class A1 Common Shares," "Class B Common Shares," "Class B1 Common Shares" and "Preferred Shares", each as designated by the Board upon original issue. The Class A Common Shares, Class A1 Common Shares, Class B Common Shares and Class B1 Common Shares are sometimes referred to herein as the "**Common Shares**" when referring to all such classes or any of them, as the context requires. In addition to the rights specifically assigned in these Bye-laws to each class of Common Shares, the Common Shares shall, subject to these Bye-laws:

(a) be entitled to such dividends as the Board may from time to time declare;

(b) in the event of a winding-up or dissolution of the Company, whether voluntary or involuntary or for the purpose of a reorganisation or otherwise or upon any distribution of capital, be entitled to the surplus assets of the Company; and

(c) generally be entitled to enjoy all of the rights attaching to shares.

4.2 The following is a statement of the additional designations and the powers, privileges and rights, and the qualifications, limitations or restrictions thereof in respect of each class of shares of the Company.

A. CLASS A COMMON SHARES

(1) General. The voting, dividend and liquidation rights of the holders of the Class A Common Shares are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares set forth herein. Except as expressly set forth in this Part A, Class A Common Shares shall have the same rights and powers of, rank equally to, share ratably with and be identical in all respects and as to all matters to Class A1 Common Shares, Class B Common Shares and Class B1 Common Shares.

(2) Voting. The holders of the Class A Common Shares are entitled to notice of and to attend all general meetings of the Company and to one (1) vote for each Class A Common Share held at all general meetings of the Company (including, for greater certainty upon the adoption of resolutions in writing in lieu of a general meeting); provided, however, that, except as otherwise required by law, holders of Class A Common Shares, as such, shall not be entitled to vote on any amendment to the Memorandum of Association or to these Bye-laws that relates solely to the terms of one or more outstanding series of Preferred Shares if the holders of such affected series are entitled, either separately or together with the holders of one or more other such series, to vote thereon pursuant to the Memorandum of Association, or pursuant to these Bye-laws or pursuant to the Act. There shall be no cumulative voting. The number of authorised Class A Common Shares may be increased or decreased (but not below the number of authorised Class A Common Shares then outstanding) by (in addition to any vote of the holders of one or more series of Preferred Shares that may be required by the terms of the Memorandum of Association or these Bye-laws) the affirmative vote of the holders of the Company's issued shares representing a majority of the voting power of the issued and outstanding shares entitled to vote.

B. CLASS B COMMON SHARES

(1) General. The voting, dividend and liquidation rights of the holders of the Class B Common Shares are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares set forth herein. Except as expressly set forth in this Part B, Class B Common Shares shall have the same rights and powers of, rank equally to, share ratably with and be identical in all respects and as to all matters to Class A Common Shares, Class A1 Common Shares and Class B1 Common Shares.

(2) Voting. The holders of the Class B Common Shares are entitled to notice of and to attend all general meetings of the Company and to ten (10) votes for each Class

B Common Share held at all general meetings of the Company (including, for greater certainty upon the adoption of resolutions in writing in lieu of a general meeting); provided, however, that, except as otherwise required by law, holders of Class B Common Shares, as such, shall not be entitled to vote on any amendment to the Memorandum of Association or to these Bye-laws that relates solely to the terms of one or more outstanding series of Preferred Shares if the holders of such affected series are entitled, either separately or together with the holders of one or more other such series, to vote thereon pursuant to the Memorandum of Association, or pursuant to these Bye-laws or pursuant to the Act. There shall be no cumulative voting. The number of authorised Class B Common Shares may be increased or decreased (but not below the number of authorised Class B Common Shares then outstanding) by (in addition to any vote of the holders of one or more series of Preferred Shares that may be required by the terms of the Memorandum of Association or these Bye-laws) the affirmative vote of the holders of the Company's issued shares representing a majority of the voting power of the issued and outstanding shares entitled to vote.

(3) Automatic Conversion. Each Class B Common Share shall automatically, without further action by the holder thereof, be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class A Common Share upon the occurrence of a Transfer, other than a Permitted Transfer, of such Class B Common Share. The Company's Register of Members shall be updated to effect such conversion immediately upon the effectiveness of such Transfer and each outstanding share certificate that, immediately prior to such Transfer, represented one or more Class B Common Shares subject to such Transfer shall, upon and after such Transfer, be deemed to represent an equal number of Class A Common Shares, without the need for surrender or exchange thereof. The Company shall, upon the request of each such holder and upon receipt of such holder's outstanding certificate, issue and deliver to such holder new certificates representing such holder's Class A Common Shares. Upon the conversion of any Class B Common Share pursuant to the foregoing provisions, the number of authorised Class B Common Shares shall be diminished by the number of Class B Common Shares that were so converted and the number of Class A Common Shares shall increase in the same amount.

For purposes of this Section B(3), the following terms shall have the following meanings:

"Family Member" shall mean with respect to any natural person who is a Qualified Shareholder, the spouse, parents, grandparents, lineal descendants, siblings and lineal descendants of siblings of such Qualified Shareholder.

"Qualified Shareholder" shall mean: (a) the registered holder of a Class B Common Share; (b) the initial registered holder of any Class B Common Shares that are originally issued by the Company pursuant to the exercise or conversion of options or

warrants or other equity awards for Class B Common Shares; (c) each natural person who Transferred Class B Common Shares or equity awards therefor (including any option or warrant exercisable or convertible into Class B Common Shares) to a Permitted Entity that is or becomes a Qualified Shareholder; and (d) a Permitted Transferee.

"Permitted Entity" shall mean with respect to a Qualified Shareholder: (a) a Permitted Trust solely for the benefit of (i) such Qualified Shareholder, (ii) one or more Family Members of such Qualified Shareholder and/or (iii) any other Permitted Entity of such Qualified Shareholder; or (b) any general partnership, limited partnership, limited liability company, corporation or other entity exclusively owned by (i) such Qualified Shareholder, (ii) one or more Family Members of such Qualified Shareholder and/or (iii) any other Permitted Entity of such Qualified Shareholder.

"Transfer" of a Class B Common Share shall mean any sale, assignment, transfer, conveyance, hypothecation or other transfer or disposition of such share or any legal or beneficial interest in such share, whether or not for value and whether voluntary or involuntary or by operation of law, including, without limitation, a transfer of such Class B Common Share to a broker or other nominee (regardless of whether there is a corresponding change in beneficial ownership).

"Permitted Transfer" shall mean, and be restricted to, any Transfer of a Class B Common Share:

(a) by a Qualified Shareholder to (i) one or more Family Members of such Qualified Shareholder, (ii) the shareholders, members, partners or other equity holders of such Qualified Shareholder or (iii) any Permitted Entity of such Qualified Shareholder; or

(b) by a Permitted Entity of a Qualified Shareholder to (i) such Qualified Shareholder or one or more Family Members of such Qualified Shareholder or (ii) any other Permitted Entity of such Qualified Shareholder.

"Permitted Transferee" shall mean a transferee of Class B Common Shares received in a Transfer that constitutes a Permitted Transfer.

"Permitted Trust" shall mean a bona fide trust where each trustee is (a) a Qualified Shareholder, (b) a Family Member or (c) a professional in the business of providing trustee services, including private professional fiduciaries, trust companies and bank trust departments.

(4) Optional Conversion.

(4.1) Each Class B Common Share shall be convertible into one (1) fully paid and non-assessable Class A Common Share at the option of the holder thereof at any time by providing written notice to the Company. Before any holder of Class B Common Shares shall be entitled to convert any of such Class B Common Shares, such holder shall surrender the certificate or certificates therefor, duly endorsed, at the principal corporate office of the Company or of any transfer agent for the Class B Common Shares, and shall give written notice to the Company at its principal corporate office, of the election to convert the same. The Company shall, as soon as practicable thereafter (and in any event, within three trading days), convert such Class B Common Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Class B Common Share shall be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class A Common Share. The Company shall issue and deliver at such office to such holder of Class B Common Shares, or to the nominee or nominees or such holder, a certificate or certificates for the number of Class A Common Shares to which such holder shall be entitled as aforesaid. Such conversion shall be deemed to have been made immediately prior to the close of business on the date of such surrender of the Class B Common Shares to be converted and the delivery to the Company of a notice of conversion, and the person or persons entitled to receive the Class A Common Shares issuable upon such conversion shall be entered in the Company's Register of Members as the record holder or holders of and treated for all purposes as the record holder or holders of such Class A Common Shares as of such date. Upon the conversion of any Class B Common Share pursuant to the foregoing provisions, the number of authorised Class B Common Shares shall be diminished by the number of Class B Common Shares that were so converted and the number of Class A Common Shares shall increase in the same amount.

(4.2) Each Class B Common Share shall be convertible into one (1) fully paid and non-assessable Class B1 Common Share at the option of the holder thereof at any time by providing written notice to the Company. Before any holder of Class B Common Shares shall be entitled to convert any of such Class B Common Shares, such holder shall surrender the certificate or certificates therefor, duly endorsed, at the principal corporate office of the Company or of any transfer agent for the Class B Common Shares, and shall give written notice to the Company at its principal corporate office, of the election to convert the same. The Company shall, as soon as practicable thereafter (and in any event, within three trading days), convert such Class B Common Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Class B Common Share shall be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class B1 Common Share. The Company shall issue and deliver at such office to such holder of Class B Common Shares, or to the nominee or nominees or such holder, a certificate or certificates for the number of Class B1 Common Shares to which such holder shall be entitled as aforesaid. Such conversion

shall be deemed to have been made immediately prior to the close of business on the date of such surrender of the Class B Common Shares to be converted and the delivery to the Company of a notice of conversion, and the person or persons entitled to receive the Class B1 Common Shares issuable upon such conversion shall be entered in the Company's Register of Members as the record holder or holders of and treated for all purposes as the record holder or holders of such Class B1 Common Shares as of such date. Upon the conversion of any Class B Common Share pursuant to the foregoing provisions, the number of authorised Class B Common Shares shall be diminished by the number of Class B Common Shares that were so converted and the number of Class B1 Common Shares shall increase in the same amount.

C. CLASS A1 COMMON SHARES

(1) General. The dividend and liquidation rights of the holders of the Class A1 Common Shares are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares set forth herein. Except as expressly set forth in this Part C, Class A1 Common Shares shall have the same rights and powers of, rank equally to, share ratably with and be identical in all respects and as to all matters to Class A Common Shares, Class B Common Shares and Class B1 Common Shares.

(2) Voting. The holders of the outstanding Class A1 Common Shares shall possess no voting power whatsoever, either general or specific. The number of authorised Class A1 Common Shares may be increased or decreased (but not below the number of shares thereof then outstanding) by (in addition to any vote of the holders of one or more series of Preferred Shares that may be required by the terms of the Memorandum of Association or these Bye-laws) the affirmative vote of the holders of the Company's issued shares representing a majority of the voting power of the issued and outstanding shares entitled to vote.

(3) Optional Conversion. Each Class A1 Common Share shall be convertible into one (1) fully paid and non-assessable Class A Common Share at the option of the holder thereof at any time by providing written notice to the Company; provided, however, that the holder of such Class A1 Common Shares shall be prohibited from converting such shares if, immediately prior to or following such conversion (or portion of such conversion thereof) the holder, together with its affiliates and any member of a Section 13(d) group, beneficially owns or would beneficially own as determined in accordance with Section 13(d) of the U.S. Securities Exchange Act of 1934, as amended, and the rules thereunder (the "**Exchange Act**") more than 4.99% (the "**Beneficial Ownership Limitation**") of the issued and outstanding Class A Common Shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act, which Beneficial Ownership Limitation may be increased, decreased or such limitation waived at such holder's election upon sixty-one (61) days' written notice to the Company. Before any holder of Class A1 Common Shares shall be entitled to convert any of such Class A1 Common

Shares, such holder shall surrender the certificate or certificates therefor, duly endorsed, at the principal corporate office of the Company or of any transfer agent for the Class A1 Common Shares, and shall give written notice to the Company at its principal corporate office, of the election to convert the same. The Company shall, as soon as practicable thereafter (and in any event, within three trading days), convert such Class A1 Common Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Class A1 Common Share shall be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class A Common Share. The Company shall issue and deliver at such office to such holder of Class A1 Common Shares, or to the nominee or nominees or such holder, a certificate or certificates for the number of Class A Common Shares to which such holder shall be entitled as aforesaid. Such conversion shall be deemed to have been made immediately prior the close of business on the date of such surrender of the Class A1 Common Shares to be converted and the delivery to the Company of a notice of conversion, and the person or persons entitled to receive the Class A Common Shares issuable upon such conversion shall be entered in the Company's Register of Members as the record holder or holders of and treated for all purposes as the record holder or holders of such Class A Common Shares as of such date. Upon the conversion of any Class A1 Common Share pursuant to the foregoing provisions, the number of authorised Class A1 Common Shares shall be diminished by the number of Class A1 Common Shares that were so converted and the number of Class A Common Shares shall increase in the same amount. Any purported delivery of any number of Class A Common Shares or any other security upon conversion of Class A1 Common Shares shall be void and have no effect to the extent, but only to the extent, that before or after such delivery, the converting holder of Class A1 Common Shares would have beneficial ownership in excess of the Beneficial Ownership Limitation. All converting holders of Class A1 Common Shares shall disclose to the Company the number of Class A Common Shares or other applicable class of equity securities that it, its affiliates or any member of a Section 13(d) group with such holder owns or has the right to acquire through the exercise or conversion of derivative securities and any limitations on exercise or conversion analogous to the limitation contained herein contemporaneously or immediately prior to submitting a notice of conversion of Class A1 Common Shares to the Company.

D. CLASS B1 COMMON SHARES

(1) General. The dividend and liquidation rights of the holders of the Class B1 Common Shares are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares set forth herein. Except as expressly set forth in this Part D, Class B1 Common Shares shall have the same rights and powers of, rank equally to, share ratably with and be identical in all respects and as to all matters to Class A Common Shares, Class A1 Common Shares and Class B Common Shares.

(2) Voting. The holders of the outstanding Class B1 Common Shares shall possess no voting power whatsoever, either general or specific. The number of authorised Class B1 Common Shares may be increased or decreased (but not below the number of shares thereof then outstanding) by (in addition to any vote of the holders of one or more series of Preferred Shares that may be required by the terms of the Memorandum of Association or these Bye-laws) the affirmative vote of the holders of the Company's issued shares representing a majority of the voting power of the issued and outstanding shares entitled to vote.

(3) Automatic Conversion. Each Class B1 Common Share shall automatically, without further action by the holder thereof, be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class A Common Share upon the occurrence of a Transfer, other than a Permitted Transfer, of such Class B1 Common Share. The Company's Register of Members shall be updated to effect such conversion immediately upon the effectiveness of such Transfer and each outstanding share certificate that, immediately prior to such Transfer, represented one or more Class B1 Common Shares subject to such Transfer shall, upon and after such Transfer, be deemed to represent an equal number of Class A Common Shares, without the need for surrender or exchange thereof. The Company shall, upon the request of each such holder and upon receipt of such holder's outstanding certificate, issue and deliver to such holder new certificates representing such holder's Class A Common Shares. Upon the conversion of any Class B1 Common Share pursuant to the foregoing provisions, the number of authorised Class B1 Common Shares shall be diminished by the number of Class B1 Common Shares that were so converted and the number of Class A Common Shares shall increase in the same amount.

For purposes of this Section D(3), the following terms shall have the following meanings:

"Family Member" shall mean with respect to any natural person who is a Qualified Shareholder, the spouse, parents, grandparents, lineal descendants, siblings and lineal descendants of siblings of such Qualified Shareholder.

"Qualified Shareholder" shall mean: (a) the registered holder of a Class B1 Common Share; (b) the initial registered holder of any Class B1 Common Shares that are originally issued by the Company pursuant to the exercise or conversion of options or warrants or other equity awards for Class B1 Common Shares; (c) each natural person who Transferred Class B1 Common Shares or equity awards therefor (including any option or warrant exercisable or convertible into Class B1 Common Shares) to a Permitted Entity that is or becomes a Qualified Shareholder; and (d) a Permitted Transferee.

"Permitted Entity" shall mean with respect to a Qualified Shareholder: (a) a Permitted Trust solely for the benefit of (i) such Qualified Shareholder, (ii) one or more Family Members of such Qualified Shareholder and/or (iii) any other Permitted Entity of such Qualified Shareholder; or (b) any general partnership, limited partnership, limited liability company, corporation or other entity exclusively owned by (i) such Qualified Shareholder, (ii) one or more Family Members of such Qualified Shareholder and/or (iii) any other Permitted Entity of such Qualified Shareholder.

"Transfer" of a Class B1 Common Share shall mean any sale, assignment, transfer, conveyance, hypothecation or other transfer or disposition of such share or any legal or beneficial interest in such share, whether or not for value and whether voluntary or involuntary or by operation of law, including, without limitation, a transfer of such Class B1 Common Share to a broker or other nominee (regardless of whether there is a corresponding change in beneficial ownership).

"Permitted Transfer" shall mean, and be restricted to, any Transfer of a Class B1 Common Share:

(a) by a Qualified Shareholder to (i) one or more Family Members of such Qualified Shareholder, (ii) the shareholders, members, partners or other equity holders of such Qualified Shareholder or (iii) any Permitted Entity of such Qualified Shareholder; or

(b) by a Permitted Entity of a Qualified Shareholder to (i) such Qualified Shareholder or one or more Family Members of such Qualified Shareholder or (ii) any other Permitted Entity of such Qualified Shareholder.

"Permitted Transferee" shall mean a transferee of Class B1 Common Shares received in a Transfer that constitutes a Permitted Transfer.

"Permitted Trust" shall mean a bona fide trust where each trustee is (a) a Qualified Shareholder, (b) a Family Member or (c) a professional in the business of providing trustee services, including private professional fiduciaries, trust companies and bank trust departments.

(4) Optional Conversion.

(4.1) Each Class B1 Common Share shall be convertible into one (1) fully paid and non-assessable Class A Common Share at the option of the holder thereof at any time by providing written notice to the Company; provided, however, that the holder of such Class B1 Common Shares shall be prohibited from converting such shares if, immediately prior to or following such conversion (or portion of such conversion thereof), the holder, together with its affiliates, and any member of a Section 13(d) group, beneficially owns or would beneficially own as determined in accordance with

Section 13(d) of the Exchange Act more than the Beneficial Ownership Limitation of the issued and outstanding Class A Common Shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act, which Beneficial Ownership Limitation may be increased, decreased or such limitation waived at such holder's election upon sixty-one (61) days' written notice to the Company. Before any holder of Class B1 Common Shares shall be entitled to convert any of such Class B1 Common Shares, such holder shall surrender the certificate or certificates therefor, duly endorsed, at the principal corporate office of the Company or of any transfer agent for the Class B1 Common Shares, and shall give written notice to the Company at its principal corporate office, of the election to convert the same. The Company shall, as soon as practicable thereafter (and in any event, within three trading days), convert such Class B1 Common Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Class B1 Common Share shall be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class A Common Share. The Company shall issue and deliver at such office to such holder of Class B1 Common Shares, or to the nominee or nominees or such holder, a certificate or certificates for the number of Class A Common Shares to which such holder shall be entitled as aforesaid. Such conversion shall be deemed to have been made immediately prior the close of business on the date of such surrender of the Class B1 Common Shares to be converted and the delivery to the Company of a notice of conversion, and the person or persons entitled to receive the Class A Common Shares issuable upon such conversion shall be entered in the Company's Register of Members as the record holder or holders of and treated for all purposes as the record holder or holders of such Class A Common Shares as of such date. Upon the conversion of any Class B1 Common Share pursuant to the foregoing provisions, the number of authorised Class B1 Common Shares shall be diminished by the number of Class B1 Common Shares that were so converted and the number of Class A Common Shares shall increase in the same amount. Any purported delivery of any number of Class A Common Shares or any other security upon conversion of Class B1 Common Shares shall be void and have no effect to the extent, but only to the extent, that before or after such delivery, the converting holder of Class B1 Common Shares would have beneficial ownership in excess of the Beneficial Ownership Limitation. All converting holders of Class B1 Common Shares shall disclose to the Company the number of Class A Common Shares or other applicable class of equity securities that it, its affiliates or any member of a Section 13(d) group with such holder owns or has the right to acquire through the exercise or conversion of derivative securities and any limitations on exercise or conversion analogous to the limitation contained herein contemporaneously or immediately prior to submitting a notice of conversion of Class B1 Common Shares to the Company.

(4.2) Each Class B1 Common Share shall be convertible into one (1) fully paid and non-assessable Class B Common Share at the option of the holder thereof at any

time by providing written notice to the Company; provided, however, that the holder of such Class B1 Common Shares shall be prohibited from converting such shares if, immediately prior to or following such conversion (or portion of such conversion thereof), the holder, together with its affiliates, and any member of a Section 13(d) group, beneficially owns or would beneficially own as determined in accordance with Section 13(d) of the Exchange Act more than the Beneficial Ownership Limitation of the issued and outstanding Class A Common Shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act, which Beneficial Ownership Limitation may be increased, decreased or such limitation waived at such holder's election upon sixty-one (61) days' written notice to the Company. Before any holder of Class B1 Common Shares shall be entitled to convert any of such Class B1 Common Shares, such holder shall surrender the certificate or certificates therefor, duly endorsed, at the principal corporate office of the Company or of any transfer agent for the Class B1 Common Shares, and shall give written notice to the Company at its principal corporate office, of the election to convert the same. The Company shall, as soon as practicable thereafter (and in any event, within three trading days), convert such Class B1 Common Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Class B1 Common Share shall be converted into and shall become (in such manner as is permitted by Bermuda law) one (1) fully paid and non-assessable Class B Common Share. The Company shall issue and deliver at such office to such holder of Class B1 Common Shares, or to the nominee or nominees or such holder, a certificate or certificates for the number of Class B Common Shares to which such holder shall be entitled as aforesaid. Such conversion shall be deemed to have been made immediately prior the close of business on the date of such surrender of the Class B1 Common Shares to be converted and the delivery to the Company of a notice of conversion, and the person or persons entitled to receive the Class B Common Shares issuable upon such conversion shall be entered in the Company's Register of Members as the record holder or holders of and treated for all purposes as the record holder or holders of such Class B Common Shares as of such date. Upon the conversion of any Class B1 Common Share pursuant to the foregoing provisions, the number of authorised Class B1 Common Shares shall be diminished by the number of Class B1 Common Shares that were so converted and the number of Class B Common Shares shall increase in the same amount. Any purported delivery of any number of Class B Common Shares or any other security upon conversion of Class B1 Common Shares shall be void and have no effect to the extent, but only to the extent, that before or after such delivery, the converting holder of Class B1 Common Shares would have beneficial ownership in excess of the Beneficial Ownership Limitation. All converting holders of Class B1 Common Shares shall disclose to the Company the number of Class A Common Shares or other applicable class of equity securities that it, its affiliates or any member of a Section 13(d) group with such holder owns or has the right to acquire through the exercise or conversion of derivative securities and any limitations on exercise or conversion analogous to the limitation contained herein

contemporaneously or immediately prior to submitting a notice of conversion of Class B1 Common Shares to the Company.

E. PREFERRED SHARES

(1) The Board is authorised to provide for the issuance of the Preferred Shares in one or more series, and to establish from time to time the number of shares to be included in each such series, and to fix the terms, including designation, powers, preferences, rights, qualifications, limitations and restrictions of the shares of each such series (and, for the avoidance of doubt, such matters and the issuance of such Preferred Shares shall not be deemed to vary the rights attached to the Common Shares or, subject to the terms of any other series of Preferred Shares, to vary the rights attached to any other series of Preferred Shares). The authority of the Board with respect to each series shall include, but not be limited to, determination of the following:

(a) the number of shares constituting that series and the distinctive designation of that series;

(b) the dividend rate on the shares of that series, whether dividends shall be cumulative and, if so, from which date or dates, and the relative rights of priority, if any, of the payment of dividends on shares of that series;

(c) whether the series shall have voting rights, in addition to the voting rights provided by law and, if so, the terms of such voting rights;

(d) whether the series shall have conversion or exchange privileges (including, without limitation, conversion into Common Shares) and, if so, the terms and conditions of such conversion or exchange, including provision for adjustment of the conversion or exchange rate in such events as the Board shall determine;

(e) whether or not the shares of that series shall be redeemable or repurchaseable and, if so, the terms and conditions of such redemption or repurchase, including the manner of selecting shares for redemption or repurchase if less than all shares are to be redeemed or repurchased, the date or dates upon or after which they shall be redeemable or repurchaseable, and the amount per share payable in case of redemption or repurchase, which amount may vary under different conditions and at different redemption or repurchase dates;

(f) whether that series shall have a sinking fund for the redemption or repurchase of shares of that series and, if so, the terms and amount of such sinking fund;

(g) the right of the shares of that series to the benefit of conditions and restrictions upon the creation of indebtedness of the Company or any subsidiary, upon the issue of any additional shares (including additional shares of such series or any

other series) and upon the payment of dividends or the making of other distributions on, and the purchase, redemption or other acquisition by the Company or any subsidiary of any issued shares of the Company;

(h) the rights of the shares of that series in the event of voluntary or involuntary liquidation, dissolution or winding up of the Company, and the relative rights of priority, if any, of payment in respect of shares of that series;

(i) the rights of holders of that series to elect or appoint directors; and

(j) any other relative participating, optional or other special rights, qualifications, limitations or restrictions of that series.

(2) Conversion. Any Preferred Shares of any series which have been redeemed (whether through the operation of a sinking fund or otherwise) or which, if convertible or exchangeable, have been converted into or exchanged for shares of any other class or classes shall have the status of authorised and unissued Preferred Shares of the same series and may be reissued as a part of the series of which they were originally a part or may be reclassified and reissued as part of a new series of Preferred Shares to be created by resolution or resolutions of the Board or as part of any other series of Preferred Shares, all subject to the conditions and the restrictions on issuance set forth in the resolution or resolutions adopted by the Board providing for the issue of any series of Preferred Shares.

4.3 At the discretion of the Board, whether or not in connection with the issuance and sale of any shares or other securities of the Company, the Company may issue securities, contracts, warrants or other instruments evidencing any shares, option rights, securities having conversion or option rights, or obligations on such terms, conditions and other provisions as are fixed by the Board including, without limiting the generality of this authority, conditions that preclude or limit any person or persons owning or offering to acquire a specified number or percentage of the issued Common Shares, other shares, option rights, securities having conversion or option rights, or obligations of the Company or transferee of the person or persons from exercising, converting, transferring or receiving the shares, option rights, securities having conversion or option rights, or obligations.

4.4 All the rights attaching to a Treasury Share shall be suspended and shall not be exercised by the Company while it holds such Treasury Share and, except where required by the Act, all Treasury Shares shall be excluded from the calculation of any percentage or fraction of the share capital, or shares, of the Company.

5. Calls on Shares

5.1 The Board may make such calls as it thinks fit upon the Members in respect of any moneys (whether in respect of nominal value or premium) unpaid on the shares allotted to or held by such Members (and not made payable at fixed times by the terms and conditions of issue) and, if a call is not paid on or before the day appointed for payment thereof, the Member may at the discretion of the Board be liable to pay the Company interest on the amount of such call at such rate as the Board may determine, from the date when such call was payable up to the actual date of payment. The Board may differentiate between the holders as to the amount of calls to be paid and the times of payment of such calls.

5.2 Any amount which, by the terms of allotment of a share, becomes payable upon issue or at any fixed date, whether on account of the nominal value of the share or by way of premium, shall for the purposes of these Bye-laws be deemed to be an amount on which a call has been duly made and payable on the date on which, by the terms of issue, the same becomes payable, and in case of non-payment all the relevant provisions of these Bye-laws as to payment of interest, costs and expenses, forfeiture or otherwise shall apply as if such amount had become payable by virtue of a duly made and notified call.

5.3 The joint holders of a share shall be jointly and severally liable to pay all calls and any interest, costs and expenses in respect thereof.

5.4 The Company may accept from any Member the whole or a part of the amount remaining unpaid on any shares held by such Member, although no part of that amount has been called up or become payable.

6. Forfeiture of Shares

6.1 If any Member fails to pay, on the day appointed for payment thereof, any call in respect of any share allotted to or held by such Member, the Board may, at any time thereafter during such time as the call remains unpaid, direct the Secretary to forward such Member a notice in writing in the form, or as near thereto as circumstances admit, of the following:

Notice of Liability to Forfeiture for Non-Payment of Call
Kiniksa Pharmaceuticals, Ltd. (the "Company")

You have failed to pay the call of [amount of call] made on [date], in respect of the [number] share(s) [number in figures] standing in your name in the Register of Members of the Company, on [date], the day appointed for payment of such call. You are hereby notified that unless you pay such call together with interest thereon at the rate of [ ] per

annum computed from the said [date] at the registered office of the Company the share(s) will be liable to be forfeited.

Dated this [date]

_____

[Signature of Secretary] By Order of the Board

6.2 If the requirements of such notice are not complied with, any such share may at any time thereafter before the payment of such call and the interest due in respect thereof be forfeited by a resolution of the Board to that effect, and such share shall thereupon become the property of the Company and may be disposed of as the Board shall determine. Without limiting the generality of the foregoing, the disposal may take place by sale, repurchase, redemption or any other method of disposal permitted by and consistent with these Bye-laws and the Act.

6.3 A Member whose share or shares have been so forfeited shall, notwithstanding such forfeiture, be liable to pay to the Company all calls owing on such share or shares at the time of the forfeiture, together with all interest due thereon and any costs and expenses incurred by the Company in connection therewith.

6.4 The Board may accept the surrender of any shares which it is in a position to forfeit on such terms and conditions as may be agreed. Subject to those terms and conditions, a surrendered share shall be treated as if it had been forfeited.

7. Share Certificates

7.1 Subject to the provisions of this Bye-law 7, every Member shall be entitled to a certificate under the common seal of the Company (or a facsimile thereof) or bearing the signature (or a facsimile thereof) of a Director or the Secretary or a person expressly authorised to sign specifying the number and, where appropriate, the class of shares held by such Member and whether the same are fully paid up and, if not, specifying the amount paid on such shares. The Board may by resolution determine, either generally or in a particular case, that any or all signatures on certificates may be printed thereon or affixed by mechanical means.

7.2 The Company shall be under no obligation to complete and deliver a share certificate unless specifically called upon to do so by the person to whom the shares have been allotted.

7.3 If any share certificate shall be proved to the satisfaction of the Board to have been worn out, lost, mislaid, or destroyed the Board may cause a new certificate to be issued and request an indemnity for the lost certificate if it sees fit.

7.4 Notwithstanding any provisions of these Bye-laws:

(a) the Board shall, subject always to the Act and any other applicable laws and regulations and the facilities and requirements of any relevant system concerned, have power to implement any arrangements it may, in its absolute discretion, think fit in relation to the evidencing of title to and transfer of uncertificated shares and to the extent such arrangements are so implemented, no provision of these Bye-laws shall apply or have effect to the extent that it is in any respect inconsistent with the holding or transfer of shares in uncertificated form; and

(b) unless otherwise determined by the Board and as permitted by the Act and any other applicable laws and regulations, no person shall be entitled to receive a certificate in respect of any share for so long as the title to that share is evidenced otherwise than by a certificate and for so long as transfers of that share may be made otherwise than by a written instrument.

8. Fractional Shares

The Company may issue its shares in fractional denominations and deal with such fractions to the same extent as its whole shares and shares in fractional denominations shall have in proportion to the respective fractions represented thereby all of the rights of whole shares including (but without limiting the generality of the foregoing) the right to vote, to receive dividends and distributions and to participate in a winding-up.

REGISTRATION OF SHARES

9. Register of Members

9.1 The Board shall cause to be kept in one or more books a Register of Members and shall enter therein the particulars required by the Act.

9.2 The Register of Members shall be open to inspection without charge at the registered office of the Company on every business day, subject to such reasonable restrictions as the Board may impose, so that not less than two hours in each business day be allowed for inspection. The Register of Members may, after notice has been given in accordance with the Act, be closed for any time or times not exceeding in the whole thirty days in each year.

10. Registered Holder Absolute Owner

The Company shall be entitled to treat the registered holder of any share as the absolute owner thereof and accordingly shall not be bound to recognise any equitable claim or other claim to, or interest in, such share on the part of any other person.

11. Transfer of Registered Shares

11.1 An instrument of transfer shall be in writing in the form of the following, or as near thereto as circumstances admit, or in such other form as the Board may accept:

Transfer of a Share or Shares
Kiniksa Pharmaceuticals, Ltd. (the "Company")

FOR VALUE RECEIVED [amount], I, [name of transferor] hereby sell, assign and transfer unto [transferee] of [address], [number] shares of the Company.

DATED this [date]

Signed by:	In the presence of:
Transferor	Witness
Signed by:	In the presence of:
Transferee	Witness

11.2 Such instrument of transfer shall be signed by (or in the case of a party that is a corporation, on behalf of) the transferor and transferee, provided that, in the case of a fully paid share, the Board may accept the instrument signed by or on behalf of the transferor alone. The transferor shall be deemed to remain the holder of such share until the same has been registered as having been transferred to the transferee in the Register of Members.

11.3 The Board may refuse to recognise any instrument of transfer unless it is accompanied by the certificate in respect of the shares to which it relates and by such other evidence

as the Board may reasonably require showing the right of the transferor to make the transfer.

11.4 The joint holders of any share may transfer such share to one or more of such joint holders, and the surviving holder or holders of any share previously held by them jointly with a deceased Member may transfer any such share to the executors or administrators of such deceased Member.

11.5 The Board may in its absolute discretion and without assigning any reason therefor refuse to register the transfer of a share which is not fully paid up. The Board shall refuse to register a transfer unless all applicable consents, authorisations and permissions of any governmental body or agency in Bermuda have been obtained. If the Board refuses to register a transfer of any share the Secretary shall, within three months after the date on which the transfer was lodged with the Company, send to the transferor and transferee notice of the refusal.

11.6 Shares may be transferred without a written instrument if transferred by an appointed agent or otherwise in accordance with the Act.

11.7 Notwithstanding anything to the contrary in these Bye-laws, shares that are listed or admitted to trading on an appointed stock exchange may be transferred in accordance with the rules and regulations of such exchange.

12. Transmission of Registered Shares

12.1 In the case of the death of a Member, the survivor or survivors where the deceased Member was a joint holder, and the legal personal representatives of the deceased Member where the deceased Member was a sole holder, shall be the only persons recognised by the Company as having any title to the deceased Member's interest in the shares. Nothing herein contained shall release the estate of a deceased joint holder from any liability in respect of any share which had been jointly held by such deceased Member with other persons. Subject to the Act, for the purpose of this Bye-law, legal personal representative means the executor or administrator of a deceased Member or such other person as the Board may, in its absolute discretion, decide as being properly authorised to deal with the shares of a deceased Member.

12.2 Any person becoming entitled to a share in consequence of the death or bankruptcy of any Member may be registered as a Member upon such evidence as the Board may deem sufficient or may elect to nominate some person to be registered as a transferee of such share, and in such case the person becoming entitled shall execute in favour of such nominee an instrument of transfer in writing in the form, or as near thereto as circumstances admit, of the following:

Transfer by a Person Becoming Entitled on Death/Bankruptcy of a Member
Kiniksa Pharmaceuticals, Ltd. (the "Company")

I/We, having become entitled in consequence of the [death/bankruptcy] of [name and address of deceased/bankrupt Member] to [number] share(s) standing in the Register of Members of the Company in the name of the said [name of deceased/bankrupt Member] instead of being registered myself/ourselves, elect to have [name of transferee] (the "Transferee") registered as a transferee of such share(s) and I/we do hereby accordingly transfer the said share(s) to the Transferee to hold the same unto the Transferee, his or her executors, administrators and assigns, subject to the conditions on which the same were held at the time of the execution hereof; and the Transferee does hereby agree to take the said share(s) subject to the same conditions.

DATED this [date]

Signed by:	In the presence of:
Transferor	Witness
Signed by:	In the presence of:
Transferee	Witness

12.3 On the presentation of the foregoing materials to the Board, accompanied by such evidence as the Board may require to prove the title of the transferor, the transferee shall be registered as a Member. Notwithstanding the foregoing, the Board shall, in any case, have the same right to decline or suspend registration as it would have had in the case of a transfer of the share by that Member before such Member's death or bankruptcy, as the case may be.

12.4 Where two or more persons are registered as joint holders of a share or shares, then in the event of the death of any joint holder or holders the remaining joint holder or holders shall be absolutely entitled to such share or shares and the Company shall recognise no claim in respect of the estate of any joint holder except in the case of the last survivor of such joint holders.

ALTERATION OF SHARE CAPITAL

13. Power to Alter Capital

13.1 The Company may if authorised by resolution of the Members increase, divide, consolidate, subdivide, change the currency denomination of, diminish or otherwise alter or reduce its share capital in any manner permitted by the Act.

13.2 Where, on any alteration or reduction of share capital, fractions of shares or some other difficulty would arise, the Board may deal with or resolve the same in such manner as it thinks fit.

14. Variation of Rights Attaching to Shares

If, at any time, the share capital is divided into different classes of shares, the rights attached to any class (unless otherwise provided by the terms of issue of the shares of that class) may, whether or not the Company is being wound-up, be varied with the consent in writing of the holders of three-fourths of the issued shares of that class or with the sanction of a resolution passed by a majority of the votes cast at a separate general meeting of the holders of the shares of the class at which meeting the necessary quorum shall be two persons at least holding or representing by proxy one-third of the issued shares of the class. The rights conferred upon the holders of the shares of any class or series issued with preferred or other rights shall not, unless otherwise expressly provided by the terms of issue of the shares of that class or series, be deemed to be varied by the creation or issue of further shares ranking *pari passu* therewith.

DIVIDENDS AND CAPITALISATION

15. Dividends

15.1 The Board may, subject to these Bye-laws and in accordance with the Act, declare a dividend to be paid to the Members, in proportion to the number of shares held by them, and such dividend may be paid in cash or wholly or partly in specie in which case the Board may fix the value for distribution in specie of any assets. No unpaid dividend shall bear interest as against the Company.

15.2 The Board may fix any date as the record date for determining the Members entitled to receive any dividend.

15.3 The Company may pay dividends in proportion to the amount paid up on each share where a larger amount is paid up on some shares than on others.

15.4 The Board may declare and make such other distributions (in cash or in specie) to the Members as may be lawfully made out of the assets of the Company. No unpaid distribution shall bear interest as against the Company.

16. Power to Set Aside Profits

The Board may, before declaring a dividend, set aside out of the surplus or profits of the Company, such amount as it thinks proper as a reserve to be used to meet contingencies or for equalising dividends or for any other purpose.

17. Method of Payment

17.1 Any dividend, interest, or other moneys payable in cash in respect of the shares may be paid by cheque or bank draft sent through the post directed to the Member at such Member's address in the Register of Members, or to such person and to such address as the Member may direct in writing, or by transfer to such account as the Member may direct in writing.

17.2 In the case of joint holders of shares, any dividend, interest or other moneys payable in cash in respect of shares may be paid by cheque or bank draft sent through the post directed to the address of the holder first named in the Register of Members, or to such person and to such address as the joint holders may direct in writing, or by transfer to such account as the joint holders may direct in writing. If two or more persons are registered as joint holders of any shares any one can give an effectual receipt for any dividend paid in respect of such shares.

17.3 The Board may deduct from the dividends or distributions payable to any Member all moneys due from such Member to the Company on account of calls or otherwise.

17.4 Any dividend and/or other moneys payable in respect of a share which has remained unclaimed for six years from the date when it became due for payment shall, if the Board so resolves, be forfeited and cease to remain owing by the Company. The payment of any unclaimed dividend or other moneys payable in respect of a share may (but need not) be paid by the Company into an account separate from the Company's own account. Such payment shall not constitute the Company a trustee in respect thereof.

17.5 The Company shall be entitled to cease sending dividend cheques and drafts by post or otherwise to a Member if those instruments have been returned undelivered to, or left uncashed by, that Member on at least two consecutive occasions or, following one such occasion, reasonable enquiries have failed to establish the Member's new address. The entitlement conferred on the Company by this Bye-law in respect of any Member shall cease if the Member claims a dividend or cashes a dividend cheque or draft.

18. Capitalisation

18.1 The Board may capitalise any amount for the time being standing to the credit of any of the Company's share premium or other reserve accounts or to the credit of the profit

and loss account or otherwise available for distribution by applying such amount in paying up unissued shares to be allotted as fully paid bonus shares pro rata (except in connection with the conversion of shares of one class to shares of another class) to the Members.

18.2 The Board may capitalise any amount for the time being standing to the credit of a reserve account or amounts otherwise available for dividend or distribution by applying such amounts in paying up in full, partly or nil paid shares of those Members who would have been entitled to such amounts if they were distributed by way of dividend or distribution.

MEETINGS OF MEMBERS

19. Annual General Meetings

An annual general meeting shall be held in each year (other than the year of incorporation) at such time and place as the president or the chairman of the Company (if any) or any two Directors or any Director and the Secretary or the Board shall appoint.

20. Special General Meetings

The president or the chairman of the Company (if any) or any two Directors or any Director and the Secretary or the Board may convene a special general meeting whenever in their judgment such a meeting is necessary.

21. Requisitioned General Meetings

The Board shall, on the requisition of Members holding at the date of the deposit of the requisition not less than one-tenth of such of the paid-up share capital of the Company as at the date of the deposit carries the right to vote at general meetings, forthwith proceed to convene a special general meeting and the provisions of the Act shall apply.

22. Notice

22.1 At least twenty days' notice of an annual general meeting shall be given to each Member entitled to attend and vote thereat, stating the date, place and time at which the meeting is to be held, that the election of Directors will take place thereat, and as far as practicable, the other business to be conducted at the meeting.

22.2 At least twenty days' notice of a special general meeting shall be given to each Member entitled to attend and vote thereat, stating the date, time, place and the general nature of the business to be considered at the meeting.

22.3 The Board may fix any date as the record date for determining the Members entitled to receive notice of and to vote at any general meeting.

22.4 A general meeting shall, notwithstanding that it is called on shorter notice than that specified in these Bye-laws, be deemed to have been properly called if it is so agreed by (i) all the Members entitled to attend and vote thereat in the case of an annual general meeting; and (ii) by a majority in number of the Members having the right to attend and vote at the meeting, being a majority together holding not less than 95% in nominal value of the shares giving a right to attend and vote thereat in the case of a special general meeting.

22.5 The accidental omission to give notice of a general meeting to, or the non-receipt of a notice of a general meeting by, any person entitled to receive notice shall not invalidate the proceedings at that meeting.

23. Giving Notice and Access

23.1 A notice may be given by the Company to a Member:

(a) by delivering it to such Member in person, in which case the notice shall be deemed to have been served upon such delivery; or

(b) by sending it by post to such Member's address in the Register of Members, in which case the notice shall be deemed to have been served seven days after the date on which it is deposited, with postage prepaid, in the mail; or

(c) by sending it by courier to such Member's address in the Register of Members, in which case the notice shall be deemed to have been served two days after the date on which it is deposited, with courier fees paid, with the courier service; or

(d) by transmitting it by electronic means (including facsimile and electronic mail, but not telephone) in accordance with such directions as may be given by such Member to the Company for such purpose, in which case the notice shall be deemed to have been served at the time that it would in the ordinary course be transmitted; or

(e) by delivering it in accordance with the provisions of the Act pertaining to delivery of electronic records by publication on a website, in which case the notice shall be deemed to have been served at the time when the requirements of the Act in that regard have been met.

23.2 Any notice required to be given to a Member shall, with respect to any shares held jointly by two or more persons, be given to whichever of such persons is named first in the Register of Members and notice so given shall be sufficient notice to all the holders of such shares.

23.3 In proving service under paragraphs 23.1(b), (c) and (d), it shall be sufficient to prove that the notice was properly addressed and prepaid, if posted or sent by courier, and

the time when it was posted, deposited with the courier, or transmitted by electronic means.

24. Postponement or Cancellation of General Meeting

The Secretary may, and on the instruction of the chairman or president of the Company or the Board, the Secretary shall, postpone or cancel any general meeting called in accordance with these Bye-laws (other than a meeting requisitioned under these Bye-laws) provided that notice of postponement or cancellation is given to the Members before the time for such meeting. Fresh notice of the date, time and place for a postponed meeting shall be given to each Member in accordance with these Bye-laws.

25. Electronic Participation and Security in Meetings

25.1 Members may participate in any general meeting by such telephonic, electronic or other communication facilities or means as permit all persons participating in the meeting to communicate with each other simultaneously and instantaneously, and participation in such a meeting shall constitute presence in person at such meeting.

25.2 The Board may, and at any general meeting, the chairman of such meeting may, make any arrangement and impose any requirement or restriction it or he considers appropriate to ensure the security of a general meeting including, without limitation, requirements for evidence of identity to be produced by those attending the meeting, the searching of their personal property and the restriction of items that may be taken into the meeting place. The Board and, at any general meeting, the chairman of such meeting are entitled to refuse entry to a person who refuses to comply with any such arrangements, requirements or restrictions.

26. Quorum at General Meetings

26.1 At any general meeting two or more persons present throughout the meeting and representing in person or by proxy a majority of the voting power of the issued and outstanding shares in the Company shall form a quorum for the transaction of business.

26.2 If within half an hour from the time appointed for the meeting a quorum is not present, then, in the case of a meeting convened on a requisition, the meeting shall be deemed cancelled and, in any other case, the meeting shall stand adjourned to the same day one week later, at the same time and place or to such other day, time or place as the Secretary may determine. Unless the meeting is adjourned to a specific date, time and place announced at the meeting being adjourned, fresh notice of the resumption of the meeting shall be given to each Member entitled to attend and vote thereat in accordance with these Bye-laws.

27. Chairman to Preside at General Meetings

Unless otherwise agreed by a majority of the voting power of the issued and outstanding shares of those attending and entitled to vote at a general meeting, the chairman of the Company, if there be one who is present, and if not the president of the Company, if there be one who is present, shall act as chairman of such meeting. In their absence a chairman of the meeting shall be appointed or elected by those present at the meeting and entitled to vote.

28. Voting on Resolutions

28.1 Subject to the Act and these Bye-laws, any question proposed for the consideration of the Members at any general meeting shall be decided by the affirmative votes of a majority of the votes cast in accordance with these Bye-laws and in the case of an equality of votes the resolution shall fail.

28.2 No Member shall be entitled to vote at a general meeting unless such Member has paid all the calls on all shares held by such Member.

28.3 At any general meeting a resolution put to the vote of the meeting shall, in the first instance, be voted upon by a show of hands and, subject to any rights or restrictions for the time being lawfully attached to any class of shares and subject to these Bye-laws, every Member present in person and every person holding a valid proxy at such meeting shall be entitled to one vote and shall cast such vote by raising his hand.

28.4 In the event that a Member participates in a general meeting by telephone, electronic or other communication facilities or means, the chairman of the meeting shall direct the manner in which such Member may cast his vote on a show of hands.

28.5 At any general meeting if an amendment is proposed to any resolution under consideration and the chairman of the meeting rules on whether or not the proposed amendment is out of order, the proceedings on the substantive resolution shall not be invalidated by any error in such ruling.

28.6 At any general meeting a declaration by the chairman of the meeting that a question proposed for consideration has, on a show of hands, been carried, or carried unanimously, or by a particular majority, or lost, and an entry to that effect in a book containing the minutes of the proceedings of the Company shall, subject to these Bye-laws, be conclusive evidence of that fact.

29. Power to Demand a Vote on a Poll

29.1 Notwithstanding the foregoing, a poll may be demanded by any of the following persons:

(a) the chairman of such meeting; or

(b) at least three Members present in person or represented by proxy; or

(c) any Member or Members present in person or represented by proxy and holding between them not less than one-tenth of the total voting rights of all the Members having the right to vote at such meeting; or

(d) any Member or Members present in person or represented by proxy holding shares in the Company conferring the right to vote at such meeting, being shares on which an aggregate sum has been paid up equal to not less than one-tenth of the total amount paid up on all such shares conferring such right.

29.2 Where a poll is demanded, subject to any rights or restrictions for the time being lawfully attached to any class of shares, every person present at such meeting shall have one vote for each share of which such person is the holder or for which such person holds a proxy and such vote shall be counted by ballot as described herein, or in the case of a general meeting at which one or more Members are present by telephone, electronic or other communication facilities or means, in such manner as the chairman of the meeting may direct and the result of such poll shall be deemed to be the resolution of the meeting at which the poll was demanded and shall replace any previous resolution upon the same matter which has been the subject of a show of hands. A person entitled to more than one vote need not use all his votes or cast all the votes he uses in the same way.

29.3 A poll demanded for the purpose of electing a chairman of the meeting or on a question of adjournment shall be taken forthwith. A poll demanded on any other question shall be taken at such time and in such manner during such meeting as the chairman (or acting chairman) of the meeting may direct. Any business other than that upon which a poll has been demanded may be conducted pending the taking of the poll.

29.4 Where a vote is taken by poll, each person physically present and entitled to vote shall be furnished with a ballot paper on which such person shall record his vote in such manner as shall be determined at the meeting having regard to the nature of the question on which the vote is taken, and each ballot paper shall be signed or initialled or otherwise marked so as to identify the voter and the registered holder in the case of a proxy. Each person present by telephone, electronic or other communication facilities or means shall cast his vote in such manner as the chairman of the meeting shall direct. At the conclusion of the poll, the ballot papers and votes cast in accordance with such directions shall be examined and counted by one or more scrutineers appointed by the Board or, in the absence of such appointment, by a committee of not less than two Members or proxy holders appointed by the chairman of the meeting for the purpose, and the result of the poll shall be declared by the chairman of the meeting.

30. Voting by Joint Holders of Shares

In the case of joint holders, the vote of the senior who tenders a vote (whether in person or by proxy) shall be accepted to the exclusion of the votes of the other joint holders, and for this purpose seniority shall be determined by the order in which the names stand in the Register of Members.

31. Instrument of Proxy

31.1 A Member may appoint a proxy by

(a) an instrument in writing in substantially the following form or such other form as the Board or the chairman of the meeting shall accept:

Proxy
Kiniksa Pharmaceuticals, Ltd. (the "Company")

I/We, [insert names here], being a Member of the Company with [number] shares, HEREBY APPOINT [name] of [address] or failing him, [name] of [address] to be my/our proxy to vote for me/us at the meeting of the Members to be held on [date] and at any adjournment thereof. [Any restrictions on voting to be inserted here.]

Signed this [date]

_____
Member(s)

or

(b) such telephonic, electronic or other means as may be approved by the Board from time to time.

31.2 The appointment of a proxy must be received by the Company at the registered office or at such other place or in such manner as is specified in the notice convening the meeting or in any instrument of proxy sent out by the Company in relation to the meeting at which the person named in the appointment proposes to vote, and appointment of a proxy which is not received in the manner so permitted shall be invalid.

31.3 A Member who is the holder of two or more shares may appoint more than one proxy to represent him and vote on his behalf in respect of different shares.

31.4 The decision of the chairman of any general meeting as to the validity of any appointment of a proxy shall be final.

32. Representation of Corporate Member

32.1 A corporation which is a Member may, by written instrument, authorise such person or persons as it thinks fit to act as its representative at any meeting and any person so authorised shall be entitled to exercise the same powers on behalf of the corporation which such person represents as that corporation could exercise if it were an individual Member, and that Member shall be deemed to be present in person at any such meeting attended by its authorised representative or representatives.

32.2 Notwithstanding the foregoing, the chairman of the meeting may accept such assurances as he thinks fit as to the right of any person to attend and vote at general meetings on behalf of a corporation which is a Member.

33. Adjournment of General Meeting

33.1 The chairman of a general meeting at which a quorum is present may, with the consent of the Members holding a majority of the voting power of the issued and outstanding shares of those Members present in person or by proxy (and shall if so directed by Members holding a majority of the voting power of the issued and outstanding shares of those Members present in person or by proxy) adjourn the meeting.

33.2 The chairman of a general meeting may adjourn the meeting to another time and place without the consent or direction of the Members if it appears to him that:

(a) it is likely to be impractical to hold or continue that meeting because of the number of Members wishing to attend who are not present; or

(b) the unruly conduct of persons attending the meeting prevents, or is likely to prevent, the orderly continuation of the business of the meeting; or

(c) an adjournment is otherwise necessary so that the business of the meeting may be properly conducted.

33.3 Unless the meeting is adjourned to a specific date, place and time announced at the meeting being adjourned, fresh notice of the date, place and time for the resumption of the adjourned meeting shall be given to each Member entitled to attend and vote thereat in accordance with these Bye-laws.

34. Written Resolutions

34.1 Subject to these Bye-laws, anything which may be done by resolution of the Company in general meeting or by resolution of a meeting of any class of the Members may be done without a meeting by written resolution in accordance with this Bye-law.

34.2 Notice of a written resolution shall be given, and a copy of the resolution shall be circulated to all Members who would be entitled to attend a meeting and vote thereon. The accidental omission to give notice to, or the non-receipt of a notice by, any Member does not invalidate the passing of a resolution.

34.3 A written resolution is passed when it is signed by (or in the case of a Member that is a corporation, on behalf of) the Members who at the date that the notice is given represent such majority of votes as would be required if the resolution was voted on at a meeting of Members at which all Members entitled to attend and vote thereat were present and voting.

34.4 A resolution in writing may be signed in any number of counterparts.

34.5 A resolution in writing made in accordance with this Bye-law is as valid as if it had been passed by the Company in general meeting or by a meeting of the relevant class of Members, as the case may be, and any reference in any Bye-law to a meeting at which a resolution is passed or to Members voting in favour of a resolution shall be construed accordingly.

34.6 A resolution in writing made in accordance with this Bye-law shall constitute minutes for the purposes of the Act.

34.7 This Bye-law shall not apply to:

(a) a resolution passed to remove an Auditor from office before the expiration of his term of office; or

(b) a resolution passed for the purpose of removing a Director before the expiration of his term of office.

34.8 For the purposes of this Bye-law, the effective date of the resolution is the date when the resolution is signed by (or in the case of a Member that is a corporation, on behalf of) the last Member whose signature results in the necessary voting majority being achieved and any reference in any Bye-law to the date of passing of a resolution is, in relation to a resolution made in accordance with this Bye-law, a reference to such date.

35. Directors Attendance at General Meetings

The Directors shall be entitled to receive notice of, attend and be heard at any general meeting.

DIRECTORS AND OFFICERS

36. Election of Directors

36.1 Only persons who are proposed or nominated in accordance with this Bye-law shall be eligible for election as Directors. Any Member or the Board may propose any person for election as a Director. Where any person, other than a Director retiring at the meeting or a person proposed for re-election or election as a Director by the Board, is to be proposed for election as a Director, notice must be given to the Company of the intention to propose him and of his willingness to serve as a Director. Where a Director is to be elected:

(a) at an annual general meeting, such notice must be given not less than 90 days nor more than 120 days before the anniversary of the last annual general meeting or, in the event the annual general meeting is called for a date that is not 30 days before or after such anniversary, the notice must be given not later than 10 days following the earlier of the date on which notice of the annual general meeting was posted to Members or the date on which public disclosure of the date of the annual general meeting was made; and

(b) at a special general meeting, such notice must be given not later than seven days following the earlier of the date on which notice of the special general meeting was posted to Members or the date on which public disclosure of the date of the special general meeting was made.

36.2 Where persons are validly proposed for re-election or election as a Director, the persons receiving the most votes (up to the number of Directors to be elected) shall be elected as Directors, and an absolute majority of the votes cast shall not be a prerequisite to the election of such Directors.

36.3 At any general meeting the Members may authorise the Board to fill any vacancy in their number left unfilled at a general meeting.

37. Number of Directors

The Board shall consist of such number of Directors being not less than five Directors and not more than such maximum number of Directors as the Board may from time to time determine.

38. Classes of Directors

The Directors shall be divided into three classes designated Class I, Class II and Class III. Each class of Directors shall consist, as nearly as possible, of one third of the total number of Directors constituting the entire Board.

39. Term of Office of Directors

At the first general meeting which is held after the date of adoption of these Bye-laws for the purpose of electing Directors, the Class I Directors shall be elected for a three year term of office, the Class II Directors shall be elected for a one year term of office and the Class III Directors shall be elected for a two year term of office. At each succeeding annual general meeting, successors to the class of Directors whose term expires at that annual general meeting shall be elected for a three year term. If the number of Directors is changed, any increase or decrease shall be apportioned among the classes so as to maintain the number of Directors in each class as nearly equal as possible, and any Director of any class elected to fill a vacancy shall hold office for a term that shall coincide with the remaining term of the other Directors of that class, but in no case shall a decrease in the number of Directors shorten the term of any Director then in office. A Director shall hold office until the annual general meeting for the year in which his term expires, subject to his office being vacated pursuant to Bye-law 41.

40. Removal of Directors

40.1 Subject to any provision to the contrary in these Bye-laws, the Members entitled to vote for the election of Directors at an annual general meeting may, at any special general meeting convened and held in accordance with these Bye-laws, remove a Director only with cause and by the affirmative vote of a majority of votes entitled to be cast by all such Members, provided that the notice of any such meeting convened for the purpose of removing a Director shall contain a statement of the intention so to do and be served on such Director not less than 14 days before the meeting and at such meeting the Director shall be entitled to be heard on the motion for such Director's removal.

40.2 If a Director is removed from the Board under this Bye-law the Members may fill the vacancy at the meeting at which such Director is removed. In the absence of such election or appointment, the Board may fill the vacancy.

40.3 For the purposes of this Bye-law, "cause" shall mean a conviction for a criminal offence involving dishonesty or engaging in conduct which brings the Director or the Company into disrepute and which results in material financial detriment to the Company.

41. Vacancy in the Office of Director

41.1 The office of Director shall be vacated if the Director:

(a) is removed from office pursuant to these Bye-laws or is prohibited from being a Director by law;

(b) is or becomes bankrupt, or makes any arrangement or composition with his creditors generally;

(c) is or becomes of unsound mind or dies; or

(d) resigns his office by notice to the Company.

41.2 The Members in general meeting or the Board shall have the power to appoint any person as a Director to fill a vacancy on the Board occurring as a result of the death, disability, disqualification or resignation of any Director or as a result of an increase in the size of the Board.

42. Remuneration of Directors

The remuneration (if any) of the Directors shall be determined by the Board and shall be deemed to accrue from day to day. The Directors may also be paid all travel, hotel and other expenses properly incurred by them (or, in the case of a director that is a corporation, by their representative or representatives) in attending and returning from Board meetings, meetings of any committee appointed by the Board or general meetings, or in connection with the business of the Company or their duties as Directors generally.

43. Defect in Appointment

All acts done in good faith by the Board, any Director, a member of a committee appointed by the Board, any person to whom the Board may have delegated any of its powers, or any person acting as a Director shall, notwithstanding that it be afterwards discovered that there was some defect in the appointment of any Director or person acting as aforesaid, or that he was, or any of them were, disqualified, be as valid as if every such person had been duly appointed and was qualified to be a Director or act in the relevant capacity.

44. Directors to Manage Business

The business of the Company shall be managed and conducted by the Board. In managing the business of the Company, the Board may exercise all such powers of the Company as are not, by the Act or by these Bye-laws, required to be exercised by the Company in general meeting.

45. Powers of the Board of Directors

The Board may:

(a) appoint, suspend, or remove any manager, secretary, clerk, agent or employee of the Company and may fix their remuneration and determine their duties;

(b) exercise all the powers of the Company to borrow money and to mortgage or charge or otherwise grant a security interest in its undertaking, property and uncalled capital, or any part thereof, and may issue debentures, debenture stock and other securities whether outright or as security for any debt, liability or obligation of the Company or any third party;

(c) appoint one or more Directors to the office of managing director or chief executive officer of the Company, who shall, subject to the control of the Board, supervise and administer all of the general business and affairs of the Company;

(d) appoint a person to act as manager of the Company's day-to-day business and may entrust to and confer upon such manager such powers and duties as it deems appropriate for the transaction or conduct of such business;

(e) by power of attorney, appoint any company, firm, person or body of persons, whether nominated directly or indirectly by the Board, to be an attorney of the Company for such purposes and with such powers, authorities and discretions (not exceeding those vested in or exercisable by the Board) and for such period and subject to such conditions as it may think fit and any such power of attorney may contain such provisions for the protection and convenience of persons dealing with any such attorney as the Board may think fit and may also authorise any such attorney to sub-delegate all or any of the powers, authorities and discretions so vested in the attorney;

(f) procure that the Company pays all expenses incurred in promoting and incorporating the Company;

(g) delegate any of its powers (including the power to sub-delegate) to a committee of one or more persons appointed by the Board which may consist partly or entirely of non-Directors, provided that every such committee shall conform to such directions as the Board shall impose on them and provided further that the meetings and proceedings of any such committee shall be governed by the provisions of these Bye-laws regulating the meetings and proceedings of the Board, so far as the same are applicable and are not superseded by directions imposed by the Board;

(h) delegate any of its powers (including the power to sub-delegate) to any person on such terms and in such manner as the Board may see fit;

(i) present any petition and make any application in connection with the liquidation or reorganisation of the Company;

(j) in connection with the issue of any share, pay such commission and brokerage as may be permitted by law; and

(k) authorise any company, firm, person or body of persons to act on behalf of the Company for any specific purpose and in connection therewith to execute any deed, agreement, document or instrument on behalf of the Company.

46. Register of Directors and Officers

The Board shall cause to be kept in one or more books at the registered office of the Company a Register of Directors and Officers and shall enter therein the particulars required by the Act.

47. Appointment of Officers

The Board may appoint such Officers (who may or may not be Directors) as the Board may determine for such terms as the Board deems fit.

48. Appointment of Secretary

The Secretary shall be appointed by the Board from time to time for such term as the Board deems fit.

49. Duties of Officers

The Officers shall have such powers and perform such duties in the management, business and affairs of the Company as may be delegated to them by the Board from time to time.

50. Remuneration of Officers

The Officers shall receive such remuneration as the Board may determine.

51. Conflicts of Interest

51.1 Any Director, or any Director's firm, partner or any company with whom any Director is associated, may act in any capacity for, be employed by or render services to the Company on such terms, including with respect to remuneration, as may be agreed between the parties. Nothing herein contained shall authorise a Director or a Director's firm, partner or company to act as Auditor to the Company.

51.2 A Director who is directly or indirectly interested in a contract or proposed contract with the Company (an "Interested Director") shall declare the nature of such interest as required by the Act.

51.3 An Interested Director who has complied with the requirements of the foregoing Bye-law may:

(a) vote in respect of such contract or proposed contract; and/or

(b) be counted in the quorum for the meeting at which the contract or proposed contract is to be voted on,

and no such contract or proposed contract shall be void or voidable by reason only that the Interested Director voted on it or was counted in the quorum of the relevant meeting and the

Interested Director shall not be liable to account to the Company for any profit realised thereby.

52. Indemnification and Exculpation of Directors and Officers

52.1 The Directors, Resident Representative, Secretary and other Officers (such term to include any person appointed to any committee by the Board) acting in relation to any of the affairs of the Company or any subsidiary thereof and the liquidator or trustees (if any) acting in relation to any of the affairs of the Company or any subsidiary thereof and every one of them (whether for the time being or formerly), and their heirs, executors and administrators (each of which an "indemnified party"), shall be indemnified and secured harmless out of the assets of the Company from and against all actions, costs, charges, losses, damages and expenses which they or any of them, their heirs, executors or administrators, shall or may incur or sustain by or by reason of any act done, concurred in or omitted in or about the execution of their duty, or supposed duty, or in their respective offices or trusts, and no indemnified party shall be answerable for the acts, receipts, neglects or defaults of the others of them or for joining in any receipts for the sake of conformity, or for any bankers or other persons with whom any moneys or effects belonging to the Company shall or may be lodged or deposited for safe custody, or for insufficiency or deficiency of any security upon which any moneys of or belonging to the Company shall be placed out on or invested, or for any other loss, misfortune or damage which may happen in the execution of their respective offices or trusts, or in relation thereto, PROVIDED THAT this indemnity shall not extend to any matter in respect of any fraud or dishonesty in relation to the Company which may attach to any of the indemnified parties. Each Member agrees to waive any claim or right of action such Member might have, whether individually or by or in the right of the Company, against any Director or Officer on account of any action taken by such Director or Officer, or the failure of such Director or Officer to take any action in the performance of his duties with or for the Company or any subsidiary thereof, PROVIDED THAT such waiver shall not extend to any matter in respect of any fraud or dishonesty in relation to the Company which may attach to such Director or Officer.

52.2 The Company may purchase and maintain insurance for the benefit of any Director or Officer against any liability incurred by him under the Act in his capacity as a Director or Officer or indemnifying such Director or Officer in respect of any loss arising or liability attaching to him by virtue of any rule of law in respect of any negligence, default, breach of duty or breach of trust of which the Director or Officer may be guilty in relation to the Company or any subsidiary thereof.

52.3 The Company may advance moneys to a Director or Officer for the costs, charges and expenses incurred by the Director or Officer in defending any civil or criminal proceedings against him, on condition that the Director or Officer shall repay the

advance if any allegation of fraud or dishonesty in relation to the Company is proved against him.

MEETINGS OF THE BOARD OF DIRECTORS

53. Board Meetings

The Board may meet for the transaction of business, adjourn and otherwise regulate its meetings as it sees fit. Subject to these Bye-laws, a resolution put to the vote at a Board meeting shall be carried by the affirmative votes of a majority of the votes cast and in the case of an equality of votes the resolution shall fail.

54. Notice of Board Meetings

A Director may, and the Secretary on the requisition of a Director shall, at any time summon a Board meeting. Notice of a Board meeting shall be deemed to be duly given to a Director if it is given to such Director verbally (including in person or by telephone) or otherwise communicated or sent to such Director by post, electronic means or other mode of representing words in a visible form at such Director's last known address or in accordance with any other instructions given by such Director to the Company for this purpose.

55. Electronic Participation in Meetings

Directors may participate in any meeting by such telephonic, electronic or other communication facilities or means as permit all persons participating in the meeting to communicate with each other simultaneously and instantaneously, and participation in such a meeting shall constitute presence in person at such meeting.

56. Representation of Corporate Director

56.1 A Director which is a corporation may, by written instrument, authorise such person or persons as it thinks fit to act as its representative at any Board meeting and any person so authorised shall be entitled to exercise the same powers on behalf of the corporation which such person represents as that corporation could exercise if it were an individual Director, and that Director shall be deemed to be present in person at any such meeting attended by its authorised representative or representatives.

56.2 Notwithstanding the foregoing, the chairman of the meeting may accept such assurances as he thinks fit as to the right of any person to attend and vote at Board meetings on behalf of a corporation which is a Director.

57. Quorum at Board Meetings

The quorum necessary for the transaction of business at a Board meeting shall be the greater of (i) a majority of the Directors at any time in office and (ii) one-third of the number of Directors established by the Board pursuant to Bye-Law 37.

58. Board to Continue in the Event of Vacancy

The Board may act notwithstanding any vacancy in its number but, if and so long as its number is reduced below the number fixed by these Bye-laws as the quorum necessary for the transaction of business at Board meetings, the continuing Directors or Director may act for the purpose of (i) summoning a general meeting; or (ii) preserving the assets of the Company.

59. Chairman to Preside

Unless otherwise agreed by a majority of the Directors attending a Board meeting, the chairman of the Company, if there be one who is present, and if not, the president of the Company, if there be one who is present, shall act as chairman at such Board meeting. In their absence a chairman of the meeting shall be appointed or elected by the Directors present at the meeting.

60. Written Resolutions

A resolution signed by (or in the case of a Director that is a corporation, on behalf of) all the Directors, which may be in counterparts, shall be as valid as if it had been passed at a Board meeting duly called and constituted, such resolution to be effective on the date on which the resolution is signed by (or in the case of a Director that is a corporation, on behalf of) the last Director.

61. Validity of Prior Acts of the Board

No regulation or alteration to these Bye-laws made by the Company in general meeting shall invalidate any prior act of the Board which would have been valid if that regulation or alteration had not been made.

CORPORATE RECORDS

62. Minutes

The Board shall cause minutes to be duly entered in books provided for the purpose:

(a) of all elections and appointments of Officers;

(b) of the names of the Directors present at each Board meeting and of any committee appointed by the Board; and

(c) of all resolutions and proceedings of general meetings of the Members, Board meetings, and meetings of committees appointed by the Board.

63. Place Where Corporate Records Kept

Minutes prepared in accordance with the Act and these Bye-laws shall be kept by the Secretary at the registered office of the Company.

64. Form and Use of Seal

64.1 The Company may adopt a seal in such form as the Board may determine. The Board may adopt one or more duplicate seals for use in or outside Bermuda.

64.2 A seal may, but need not, be affixed to any deed, instrument or document, and if the seal is to be affixed thereto, it shall be attested by the signature of (i) any Director, or (ii) any Officer, or (iii) the Secretary, or (iv) any person authorised by the Board for that purpose.

64.3 A Resident Representative may, but need not, affix the seal of the Company to certify the authenticity of any copies of documents.

ACCOUNTS

65. Records of Account

65.1 The Board shall cause to be kept proper records of account with respect to all transactions of the Company and in particular with respect to:

(a) all amounts of money received and expended by the Company and the matters in respect of which the receipt and expenditure relates;

(b) all sales and purchases of goods by the Company; and

(c) all assets and liabilities of the Company.

65.2 Such records of account shall be kept at the registered office of the Company or, subject to the Act, at such other place as the Board thinks fit and shall be available for inspection by the Directors during normal business hours.

65.3 Such records of account shall be retained for a minimum period of five years from the date on which they are prepared.

66. Financial Year End

The financial year end of the Company may be determined by resolution of the Board and failing such resolution shall be 31st December in each year.

AUDITS

67. Annual Audit

Subject to any rights to waive laying of accounts or appointment of an Auditor pursuant to the Act, the accounts of the Company shall be audited at least once in every year.

68. Appointment of Auditor

68.1 Subject to the Act, the Members shall appoint an auditor to the Company to hold office for such term as the Members deem fit or until a successor is appointed.

68.2 The Auditor may be a Member but no Director, Officer or employee of the Company shall, during his continuance in office, be eligible to act as an Auditor of the Company.

69. Remuneration of Auditor

69.1 The remuneration of an Auditor appointed by the Members shall be fixed by the Company in general meeting or in such manner as the Members may determine.

69.2 The remuneration of an Auditor appointed by the Board to fill a casual vacancy in accordance with these Bye-laws shall be fixed by the Board.

70. Duties of Auditor

70.1 The financial statements provided for by these Bye-laws shall be audited by the Auditor in accordance with generally accepted auditing standards. The Auditor shall make a written report thereon in accordance with generally accepted auditing standards.

70.2 The generally accepted auditing standards referred to in this Bye-law may be those of a country or jurisdiction other than Bermuda or such other generally accepted auditing standards as may be provided for in the Act. If so, the financial statements and the report of the Auditor shall identify the generally accepted auditing standards used.

71. Access to Records

The Auditor shall at all reasonable times have access to all books kept by the Company and to all accounts and vouchers relating thereto, and the Auditor may call on the Directors or Officers for any information in their possession relating to the books or affairs of the Company.

72. Financial Statements and the Auditor's Report

72.1 Subject to the following bye-law, the financial statements and/or the auditor's report as required by the Act shall

(a) be laid before the Members at the annual general meeting; or

(b) be received, accepted, adopted or approved by the Members by written resolution passed in accordance with these Bye-laws.

72.2 If all Members and Directors shall agree, either in writing or at a meeting, that in respect of a particular interval no financial statements and/or auditor's report thereon need be made available to the Members, and/or that no auditor shall be appointed then there shall be no obligation on the Company to do so.

73. Vacancy in the Office of Auditor

The Board may fill any casual vacancy in the office of the auditor.

BUSINESS COMBINATIONS

74. Business Combinations

74.1 (a) Any Business Combination with any Interested Shareholder within a period of three years following the time of the transaction in which the person became an Interested Shareholder must be approved by the Board and authorised at an annual or special general meeting, by the affirmative vote of a majority of the voting power of the issued and outstanding voting shares of the Company that are not owned by the Interested Shareholder unless:

(i) prior to the time that the person became an Interested Shareholder, the Board approved either the Business Combination or the transaction which resulted in the person becoming an Interested Shareholder;

(ii) upon consummation of the transaction which resulted in the person becoming an Interested Shareholder, the Interested Shareholder owned at least 85% of the voting power of the issued and outstanding voting shares of the Company at the time the transaction commenced, excluding for the purposes of determining the number of shares issued and outstanding those shares owned (i) by persons who are Directors and also Officers and (ii) employee share plans in which employee participants do not have the right to determine whether shares held subject to the plan will be tendered in a tender or exchange offer; or

(iii) following the date of the consummation of the transaction which resulted in the person becoming an Interested Shareholder, the Business Combination is approved by the Board and authorized at an annual or special general meeting, by the affirmative vote of at least $66^{2}/_{3}$% of the voting power of the issued and outstanding voting shares of the Company that are not owned by the Interested Shareholder.

(b) The restrictions contained in this Bye-law 74.1 shall not apply if:

(i) a Member becomes an Interested Shareholder inadvertently and (i) as soon as practicable divests itself of ownership of sufficient shares so that the Member ceases to be an Interested Shareholder; and (ii) would not, at any time within the three-year period immediately prior to a Business Combination between the Company and such Member, have been an Interested Shareholder but for the inadvertent acquisition of ownership; or

(ii) the Business Combination is proposed prior to the consummation or abandonment of, and subsequent to the earlier of the public announcement or the notice required hereunder of, a proposed transaction which (i) constitutes one of the transactions described in the following sentence; (ii) is with or by a person who either was not an Interested Shareholder during the previous three years or who became an Interested Shareholder with the approval of the Board; and (iii) is approved or not opposed by a majority of the members of the Board then in office who were Directors prior to any person becoming an Interested Shareholder during the previous three years or were recommended for election or elected to succeed such Directors by resolution of the Board approved by a majority of such Directors. The proposed transactions referred to in the preceding sentence are limited to:

(a) a merger, amalgamation or consolidation of the Company (except a merger or amalgamation in respect of which, pursuant to the Act, no vote of the Members is required);

(b) a sale, lease, exchange, mortgage, pledge, transfer or other disposition (in one transaction or a series of transactions), whether as part of a dissolution or otherwise, of assets of the Company or of any entity directly or indirectly wholly-owned or majority-owned by the Company (other than to the Company or any entity directly or indirectly wholly-owned by the Company) having an aggregate market value equal to 50% or more of either the aggregate market value of all of the assets of the Company determined on a consolidated basis or the aggregate market value of all the issued and outstanding shares of the Company; or

(c) a proposed tender or exchange offer for 50% or more of the voting power of the issued and outstanding voting shares of the Company.

(c) For the purpose of this Bye-law 74 only, the term:

(i) "affiliate" means a person that directly, or indirectly through one or more intermediaries, controls, or is controlled by, or is under common control with, another person;

(ii) "associate", when used to indicate a relationship with any person, means: (i) any company, partnership, unincorporated association or other entity of which such person is a director, officer or partner or is, directly or indirectly, the owner of 20% or more of any class of voting shares; (ii) any trust or other estate in which such person has at least a 20% beneficial interest or as to which such person serves as trustee or in a similar fiduciary capacity; and (iii) any relative or spouse of such person, or any relative of such spouse, who has the same residence as such person;

(iii) "Business Combination", when used in reference to the Company and any Interested Shareholder of the Company, means:

(a) any merger, amalgamation or consolidation of the Company or any entity directly or indirectly wholly-owned or majority-owned by the Company, wherever incorporated, with (A) the Interested Shareholder or any of its affiliates, or (B) with any other company, partnership, unincorporated association or other entity if the merger, amalgamation or consolidation is caused by the Interested Shareholder;

(b) any sale, lease, exchange, mortgage, pledge, transfer or other disposition (in one transaction or a series of transactions), except proportionately as a shareholder of the Company, to or with the Interested Shareholder, whether as part of a dissolution or otherwise, of assets of the Company or of any entity directly or indirectly wholly-owned or majority-owned by the Company which assets have an aggregate market value equal to 10% or more of either the aggregate market value of all the assets of the Company determined on a consolidated basis or the aggregate market value of all the issued and outstanding shares of the Company;

(c) any transaction which results in the issuance or transfer by the Company or by any entity directly or indirectly wholly-owned or majority-owned by the Company of any shares of the Company, or any share of such entity, to the Interested Shareholder, except: (A) pursuant to the exercise, exchange or

conversion of securities exercisable for, exchangeable for or convertible into shares of the Company, or shares of any such entity, which securities were issued and outstanding prior to the time that the Interested Shareholder became such; (B) pursuant to a dividend or distribution paid or made, or the exercise, exchange or conversion of securities exercisable for, exchangeable for or convertible into shares of the Company, or shares of any such entity, which security is distributed, pro rata to all holders of a class or series of shares subsequent to the time the Interested Shareholder became such; (C) pursuant to an exchange offer by the Company to purchase shares made on the same terms to all holders of such shares; or (D) any issuance or transfer of shares by the Company; provided however, that in no case under items (B)-(D) of this subparagraph shall there be an increase in the Interested Shareholder's proportionate share of any class or series of shares;

(d) any transaction involving the Company or any entity directly or indirectly wholly-owned or majority-owned by the Company which has the effect, directly or indirectly, of increasing the proportionate share of any class or series of shares, or securities convertible into any class or series of shares of the Company, or shares of any such entity, or securities convertible into such shares, which is owned by the Interested Shareholder, except as a result of immaterial changes due to fractional share adjustments or as a result of any repurchase or redemption of any shares not caused, directly or indirectly, by the Interested Shareholder; or

(e) any receipt by the Interested Shareholder of the benefit, directly or indirectly (except proportionately as a shareholder of the Company), of any loans, advances, guarantees, pledges or other financial benefits (other than those expressly permitted in subparagraphs (a)-(d) of this paragraph) provided by or through the Company or any entity directly or indirectly wholly-owned or majority-owned by the Company;

(iv) "control", including the terms "controlling", "controlled by" and "under common control with", means the possession, directly or indirectly, of the power to direct or cause the direction of the management and policies of a person, whether through the ownership of voting shares, by contract or otherwise. A person who is the owner of 20% or more of the voting power of the issued and outstanding voting shares of any

company, partnership, unincorporated association or other entity shall be presumed to have control of such entity, in the absence of proof by a preponderance of the evidence to the contrary; provided that notwithstanding the foregoing, such presumption of control shall not apply where such person holds voting shares, in good faith and not for the purpose of circumventing this provision, as an agent, bank, broker, nominee, custodian or trustee for one or more owners who do not individually or as a group have control of such entity;

(v) "Interested Shareholder" means any person (other than the Company and any entity directly or indirectly wholly-owned or majority-owned by the Company) that (i) is the owner of 15% or more of the issued and outstanding voting shares of the Company, (ii) is an affiliate or associate of the Company and was the owner of 15% or more of the issued and outstanding voting shares of the Company at any time within the three-year period immediately prior to the date on which it is sought to be determined whether such person is an Interested Shareholder or (iii) is an affiliate or associate of any person listed in (i) or (ii) above; provided, however, that the term "Interested Shareholder" shall not include any person whose ownership of shares in excess of the 15% limitation set forth herein is the result of action taken solely by the Company unless such person referred to in this proviso acquires additional voting shares of the Company otherwise than as a result of further corporate action not caused, directly or indirectly, by such person. For the purpose of determining whether a person is an Interested Shareholder, the voting shares of the Company deemed to be issued and outstanding shall include voting shares deemed to be owned by the person through application of paragraph (8) below, but shall not include any other unissued shares which may be issuable pursuant to any agreement, arrangement or understanding, or upon exercise of conversion rights, warrants or options, or otherwise;

(vi) "person" means any individual, company, partnership, unincorporated association or other entity;

(vii) "voting shares" means, with respect to any company, shares of any class or series entitled to vote generally in the election of directors, provided that, when used in reference to a vote to approve a merger or amalgamation of the Company which the Act requires to be approved by the Members, such term includes any shares entitled to vote on such matter pursuant to the Act, whether or not they are otherwise entitled to vote and, with respect to any entity that is not a company, any equity interest entitled to vote generally in the election of the governing body

of such entity; and references to percentages of "voting shares" shall be read as references to shares carrying such percentages of votes;

(viii) "owner", including the terms "own" and "owned", when used with respect to any shares, means a person that individually or with or through any of its affiliates or associates:

(a) beneficially owns such shares, directly or indirectly; or

(b) has (A) the right to acquire such shares (whether such right is exercisable immediately or only after the passage of time) pursuant to any agreement, arrangement or understanding, or upon the exercise of conversion rights, exchange rights, warrants or options, or otherwise; provided, however, that a person shall not be deemed the owner of shares tendered pursuant to a tender or exchange offer made by such person or any of such person's affiliates or associates until such tendered shares are accepted for purchase or exchange; or (B) the right to vote such shares pursuant to any agreement, arrangement or understanding; provided, however, that a person shall not be deemed the owner of any shares because of such person's right to vote such shares if the agreement, arrangement or understanding to vote such shares arises solely from a revocable proxy or consent given in response to a proxy or consent solicitation made to 10 or more persons; or

(c) has any agreement, arrangement or understanding for the purpose of acquiring, holding, voting (except voting pursuant to a revocable proxy or consent as described in item (B) of subparagraph (b) of this paragraph), or disposing of such shares with any other person that beneficially owns, or whose affiliates or associates beneficially own, directly or indirectly, such shares.

74.2 In respect of any Business Combination to which the restrictions contained in Bye-law 74.1 do not apply but which the Act requires to be approved by the Members:

(a) where such Business Combination has been approved by the Board, the necessary general meeting quorum and Members' approval shall be as set out in Bye-laws 26 and 28 respectively; and

(b) where such Business Combination has not been approved by the Board, the necessary Members' approval shall require the affirmative vote of at least $66^{2}/_{3}$% of the voting power of the issued and outstanding voting shares of the Company.

74.3 In respect of any merger or amalgamation which is not a Business Combination but which the Act requires to be approved by the Members:

(a) where such merger or amalgamation has been approved by the Board, the necessary general meeting quorum and Members' approval shall be as set out in Bye-laws 26 and 28 respectively; and

(b) where such merger or amalgamation has not been approved by the Board, the necessary Members' approval shall require the affirmative vote of at least 66 2/3% of the voting power of the issued and outstanding voting shares of the Company.

74.4 The Board shall ensure that the bye-laws or other constitutional documents of each entity wholly-owned or majority-owned by the Company shall contain any provisions necessary to ensure that the intent of Bye-law 74.1, as it relates to the actions of such entities, is achieved.

VOLUNTARY WINDING-UP AND DISSOLUTION

75. Winding-Up

If the Company shall be wound up the liquidator may, with the sanction of a resolution of the Members, divide amongst the Members in specie or in kind the whole or any part of the assets of the Company (whether they shall consist of property of the same kind or not) and may, for such purpose, set such value as he deems fair upon any property to be divided as aforesaid and may determine how such division shall be carried out as between the Members or different classes of Members. The liquidator may, with the like sanction, vest the whole or any part of such assets in the trustees upon such trusts for the benefit of the Members as the liquidator shall think fit, but so that no Member shall be compelled to accept any shares or other securities or assets whereon there is any liability.

CHANGES TO CONSTITUTION

76. Changes to Bye-laws

76.1 Subject to Bye-law 76.2, no Bye-law may be rescinded, altered or amended and no new Bye-law may be made save in accordance with the Act and until the same has been approved by a resolution of the Board and by a resolution of the Members.

76.2 Bye-laws 36, 37, 38, 39, 40, 74 and 76 may not be rescinded, altered or amended and no new Bye-law may be made which would have the effect of rescinding, altering or amending the provisions of such Bye-laws, until the same has been approved by a resolution of the Board including the affirmative vote of not less than 66% of the Directors then in office and by a resolution of the Members including the affirmative

vote of shares carrying not less than 66% of the voting power of the issued and outstanding shares.

77. Discontinuance

The Board may exercise all the powers of the Company to discontinue the Company to a jurisdiction outside Bermuda pursuant to the Act.

78. Default Jurisdiction

In the event that any dispute arises concerning the Act or out of or in connection with these Bye-laws, including any question regarding the existence and scope of any Bye-law and/or whether there has been a breach of the Act or these Bye-laws by any Officer or Director (whether or not such a claim is brought in the name of a shareholder or in the name of the Company), and unless the Company consents in writing to the selection of an alternative jurisdiction, any such dispute shall be subject to the jurisdiction of the Supreme Court of Bermuda. If any provision or provisions of this Bye-Law 78 shall be held to be invalid, illegal or unenforceable as applied to any person or entity or circumstance for any reason whatsoever, then, to the fullest extent permitted by law, the validity, legality and enforceability of such provisions in any other circumstance and of the remaining provisions of this Bye-Law 78 (including, without limitation, each portion of any sentence of this Bye-Law 78 containing any such provision held to be invalid, illegal or unenforceable that is not itself held to be invalid, illegal or unenforceable) and the application of such provision to other persons or entities and circumstances shall not in any way be affected or impaired thereby.


CLASS A COMMON SHARES
NUMBER
KPL
KINIKSA
CLASS A COMMON SHARES
SHARES
KINIKSA PHARMACEUTICALS, LTD.
INCORPORATED UNDER THE LAWS OF BERMUDA
CUSIP
SEE REVERSE FOR CERTAIN DEFINITIONS
THIS CERTIFIES THAT
SPECIMEN
IS THE RECORD HOLDER OF
FULLY PAID AND NON-ASSESSABLE CLASS A COMMON SHARES, US $.000273235 PAR VALUE PER SHARE, OF
KINIKSA PHARMACEUTICALS, LTD.
held subject to the memorandum of association and bye-laws of the Company and transferable in accordance therewith. This certificate is not valid until countersigned by the Transfer Agent and registered by the Registrar.
Witness the facsimile seal of the Company and the facsimile signature of its duly authorized officer.
Dated:
Kiniksa Pharmaceuticals, Ltd
BERMUDA
2018
CHAIRMAN AND CHIEF EXECUTIVE OFFICER
AUTHORIZED SIGNATURE
COUNTERSIGNED AND REGISTERED:
AMERICAN STOCK TRANSFER & TRUST COMPANY, LLC
(Brooklyn, NY)
BY
TRANSFER AGENT AND REGISTRAR
SECURITY COLUMBIAN UNITED STATES BANKNOTE COMPANY 1960

The following abbreviations, when used in the inscription on the face of this certificate, shall be construed as though they were written out in full according to applicable laws or regulations:

TEN COM — as tenants in common
TEN ENT — as tenants by the entireties
JT TEN — as joint tenants with rights of survivorship and not as tenants in common

UNIF GIFT MIN ACT — _____ (Cust) Custodian _____ (Minor)
under Uniform Gifts to Minors
Act _____ (State)

Additional abbreviations may also be used though not in the above list.

For value received, _____ hereby sell, assign and transfer unto

PLEASE INSERT SOCIAL SECURITY OR OTHER IDENTIFYING NUMBER OF ASSIGNEE

_____

_____

(PLEASE PRINT NAME AND ADDRESS, INCLUDING ZIP CODE, OF ASSIGNEE)

_____ Shares of Class A Shares represented by the within Certificate, and do hereby irrevocably constitute and appoint

_____ Attorney to transfer the said stock on the books of the within-named Company with full power of substitution in the premises.

Dated:

X _____

X _____

NOTICE: THE SIGNATURE TO THIS ASSIGNMENT MUST CORRESPOND WITH THE NAME AS WRITTEN UPON THE FACE OF THE CERTIFICATE IN EVERY PARTICULAR WITHOUT ALTERATION OR ENLARGEMENT OR ANY CHANGE WHATEVER.

SIGNATURE GUARANTEED:

_____

ALL GUARANTEES MUST BE MADE BY A FINANCIAL INSTITUTION (SUCH AS A BANK OR BROKER) WHICH IS A PARTICIPANT IN THE SECURITIES TRANSFER AGENTS MEDALLION PROGRAM ("STAMP"), THE NEW YORK STOCK EXCHANGE, INC. MEDALLION SIGNATURE PROGRAM ("MSP"), OR THE STOCK EXCHANGES MEDALLION PROGRAM ("SEMP") PURSUANT TO S.E.C. RULE 17Ad-15, AND MUST NOT BE DATED, GUARANTEES BY A NOTARY PUBLIC ARE NOT ACCEPTABLE.

Exhibit 5.1

14 May 2018

Matter No.:883185
Doc Ref: 104160250
+441 298 7859
chiara.nannini@conyersdill.com

Kiniksa Pharmaceuticals, Ltd.
Clarendon House
2 Church Street
Hamilton HM 11
Bermuda

Dear Sirs,

Re: **Kiniksa Pharmaceuticals, Ltd. (the "Company")**

We have acted as special Bermuda legal counsel to the Company in connection with a registration statement on form S-1 (Registration No. 333-224488) filed with the U.S. Securities and Exchange Commission (the "Commission") on 14 May 2018 (the "Registration Statement", which term does not include any other document or agreement whether or not specifically referred to therein or attached as an exhibit or schedule thereto) relating to the registration under the U.S. Securities Act of 1933, as amended, (the "Securities Act") of 7,000,000 Class A common shares of the Company, par value US$0.000273235 each being offered by the Company (the "IPO Shares") together with an additional 1,050,000 Class A common shares, par value US$0.000273235 each subject to an over-allotment option granted to the underwriters by the Company (the "Over-Allotment Shares") and any Class A common shares, par value US$0.000273235 that may be sold pursuant to an additional registration statement filed pursuant to Rule 462(b) under the Securities Act, if any (the "Additional Shares" and together with the Over-Allotment Shares and the IPO Shares, the "Class A Common Shares").

For the purposes of giving this opinion, we have examined a copy of the Registration Statement. We have also reviewed the memorandum of association and the bye-laws of the Company, each certified by the Secretary of the Company on 14 May 2018, minutes of a meeting of its directors held on 10 May 2018 (the "Minutes") and written resolutions adopted on 11 May 2018 by the shareholders of the Company (together, the Minutes and the written resolutions adopted by the shareholders of the Company, the "Resolutions") and

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such other documents and made such enquiries as to questions of law as we have deemed necessary in order to render the opinion set forth below.

We have assumed (a) the genuineness and authenticity of all signatures and the conformity to the originals of all copies (whether or not certified) examined by us and the authenticity and completeness of the originals from which such copies were taken, (b) that where a document has been examined by us in draft form, it will be or has been executed and/or filed in the form of that draft, and where a number of drafts of a document have been examined by us all changes thereto have been marked or otherwise drawn to our attention, (c) the accuracy and completeness of all factual representations made in the Registration Statement and other documents reviewed by us, (d) that the Resolutions were passed at one or more duly convened, constituted and quorate meetings, or by written resolutions, remain in full force and effect and have not been rescinded or amended, (e) that there is no provision of the law of any jurisdiction, other than Bermuda, which would have any implication in relation to the opinions expressed herein, (f) that upon issue of any shares to be sold by the Company, the Company will receive consideration for the full issue price thereof which shall be equal to at least the par value thereof. "Non-assessability" is not a legal concept under Bermuda law, but when we described the Class A Common Shares herein as being "non-assessable" we mean that no further sums are payable with respect to the issue of such shares.

We have made no investigation of and express no opinion in relation to the laws of any jurisdiction other than Bermuda. This opinion is to be governed by and construed in accordance with the laws of Bermuda and is limited to and is given on the basis of the current law and practice in Bermuda. This opinion is issued solely for the purposes of the filing of the Registration Statement and the offering of the Class A Common Shares by the Company and is not to be relied upon in respect of any other matter.

On the basis of and subject to the foregoing, we are of the opinion that:

1. The Company is duly incorporated and existing under the laws of Bermuda in good standing (meaning solely that it has not failed to make any filing with any Bermuda government authority or to pay any Bermuda government fees or tax which would make it liable to be struck off the Register of Companies and thereby cease to exist under the laws of Bermuda).

2. When issued and paid for as contemplated by the Registration Statement, the Class A Common Shares will be validly issued, fully paid and non-assessable.

We hereby consent to the filing of this opinion as an exhibit to the Registration Statement and to the references to our firm under the caption "Legal Matters" in the prospectus forming a part of the Registration Statement. In giving this consent, we do not hereby admit that we are experts within the meaning of Section 11 of the Securities Act or that we are

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within the category of persons whose consent is required under Section 7 of the Securities Act or the Rules and Regulations of the Commission promulgated thereunder.

Yours faithfully,

/s/ Conyers Dill & Pearman Limited

Conyers Dill & Pearman Limited

KINIKSA PHARMACEUTICALS, LTD.
2018 INCENTIVE AWARD PLAN

ARTICLE I.
PURPOSE

The Plan's purpose is to enhance the Company's ability to attract, retain and motivate persons who make (or are expected to make) important contributions to the Company by providing these individuals with equity ownership opportunities. Capitalized terms used in the Plan are defined in Article XI.

ARTICLE II.
ELIGIBILITY

Service Providers are eligible to be granted Awards under the Plan, subject to the limitations described herein.

ARTICLE III.
ADMINISTRATION AND DELEGATION

3.1 Administration. The Plan is administered by the Administrator. The Administrator has authority to determine which Service Providers receive Awards, grant Awards and set Award terms and conditions, subject to the conditions and limitations in the Plan. The Administrator also has the authority to take all actions and make all determinations under the Plan, to interpret the Plan and Award Agreements and to adopt, amend and repeal Plan administrative rules, guidelines and practices as it deems advisable. The Administrator may correct defects and ambiguities, supply omissions and reconcile inconsistencies in the Plan or any Award as it deems necessary or appropriate to administer the Plan and any Awards. The Administrator's determinations under the Plan are in its sole discretion and will be final and binding on all persons having or claiming any interest in the Plan or any Award.

3.2 Appointment of Committees. To the extent Applicable Laws permit, the Board may delegate any or all of its powers under the Plan to one or more Committees or officers of the Company or any of its Subsidiaries. The Board may abolish any Committee or re-vest in itself any previously delegated authority at any time.

ARTICLE IV.
SHARES AVAILABLE FOR AWARDS

4.1 Number of Shares. Subject to adjustment under Article VIII and the terms of this Article IV, Awards may be made under the Plan covering up to the Overall Share Limit. As of the Plan's effective date under Section 10.3, the Company will cease granting awards under the Prior Plans; however, Prior Plan Awards will remain subject to the terms of the applicable Prior Plan. Shares issued under the Plan may consist of authorized but unissued Shares, Shares purchased on the open market or Shares held in treasury.

4.2 Share Recycling. If all or any part of an Award or Prior Plan Award expires, lapses or is terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited, in any case, in a manner that results in the Company acquiring Shares covered by the Award or Prior Plan Award at a price not greater than the price (as adjusted to reflect any Equity Restructuring) paid by the Participant for such Shares or not issuing any Shares covered by the Award or Prior Plan Award, the unused Shares covered by the Award or Prior Plan Award will, as applicable, become or again be available

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for Award grants under the Plan. Further, Shares delivered (either by actual delivery or attestation) to the Company by a Participant to satisfy the applicable exercise or purchase price of an Award or Prior Plan Award and/or to satisfy any applicable tax withholding obligation, will, as applicable, become or again be available for Award grants under the Plan. The payment of Dividend Equivalents in cash in conjunction with any outstanding Awards shall not count against the Overall Share Limit.

4.3 Incentive Stock Option Limitations. Notwithstanding anything to the contrary herein, no more than 27,915,000 Shares may be issued pursuant to the exercise of Incentive Stock Options.

4.4 Substitute Awards. In connection with an entity's amalgamation, merger or consolidation with the Company or the Company's acquisition of an entity's property or shares, the Administrator may grant Awards in substitution for any options or other shares or share-based awards granted before such amalgamation, merger or consolidation by such entity or its affiliate. Substitute Awards may be granted on such terms as the Administrator deems appropriate, notwithstanding limitations on Awards in the Plan. Substitute Awards will not count against the Overall Share Limit (nor shall Shares subject to a Substitute Award be added to the Shares available for Awards under the Plan as provided above), except that Shares acquired by exercise of substitute Incentive Stock Options will count against the maximum number of Shares that may be issued pursuant to the exercise of Incentive Stock Options under the Plan. Additionally, in the event that a company acquired by the Company or any Subsidiary or with which the Company or any Subsidiary combines has shares available under a pre-existing plan approved by shareholders and not adopted in contemplation of such acquisition or combination, the shares available for grant pursuant to the terms of such pre-existing plan (as adjusted, to the extent appropriate, using the exchange ratio or other adjustment or valuation ratio or formula used in such acquisition or combination to determine the consideration payable to the equity holders of the entities party to such acquisition or combination) may be used for Awards under the Plan and shall not reduce the Shares authorized for grant under the Plan (and Shares subject to such Awards shall not be added to the Shares available for Awards under the Plan as provided above); provided that Awards using such available shares shall not be made after the date awards or grants could have been made under the terms of the pre-existing plan, absent the acquisition or combination, and shall only be made to individuals who were not Employees or Directors prior to such acquisition or combination.

ARTICLE V.
SHARE OPTIONS AND SHARE APPRECIATION RIGHTS

5.1 General. The Administrator may grant Options or Share Appreciation Rights to Service Providers subject to the limitations in the Plan, including any limitations in the Plan that apply to Incentive Stock Options. The Administrator will determine the number of Shares covered by each Option and Share Appreciation Right, the exercise price of each Option and Share Appreciation Right, the vesting conditions of each Option and Share Appreciation Right, and the conditions and limitations applicable to the exercise of each Option and Share Appreciation Right. A Share Appreciation Right will entitle the Participant (or other person entitled to exercise the Share Appreciation Right) to receive from the Company upon exercise of the exercisable portion of the Share Appreciation Right an amount determined by multiplying the excess, if any, of the Fair Market Value of one Share on the date of exercise over the exercise price per Share of the Share Appreciation Right by the number of Shares with respect to which the Share Appreciation Right is exercised, subject to any limitations of the Plan or that the Administrator may impose and payable in cash, Shares valued at Fair Market Value or a combination of the two as the Administrator may determine or provide in the Award Agreement.

5.2 Exercise Price. The Administrator will establish each Option's and Share Appreciation Right's exercise price and specify the exercise price in the Award Agreement. The exercise price will not

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be less than 100% of the Fair Market Value or par value per Share, whichever is greater, on the grant date of the Option or Share Appreciation Right.

5.3 Duration. Each Option or Share Appreciation Right will be exercisable at such times and as specified in the Award Agreement, provided that the term of an Option or Share Appreciation Right will not exceed ten years. Notwithstanding the foregoing and unless determined otherwise by the Company, in the event that on the last business day of the term of an Option or Share Appreciation Right (other than an Incentive Stock Option) (i) the exercise of the Option or Share Appreciation Right is prohibited by Applicable Law, as determined by the Company, or (ii) Shares may not be purchased or sold by the applicable Participant due to any Company insider trading policy (including blackout periods) or a "lock-up" agreement undertaken in connection with an issuance of securities by the Company, the term of the Option or Share Appreciation Right shall be extended until the date that is thirty (30) days after the end of the legal prohibition, black-out period or lock-up agreement, as determined by the Company; provided, however, in no event shall the extension last beyond the ten year term of the applicable Option or Share Appreciation Right. Notwithstanding the foregoing, if the Participant, prior to the end of the term of an Option or Share Appreciation Right, violates the non-competition, non-solicitation, confidentiality or other similar restrictive covenant provisions of any employment contract, confidentiality and nondisclosure agreement or other agreement between the Participant and the Company or any of its Subsidiaries, the right of the Participant and the Participant's transferees to exercise any Option or Share Appreciation Right issued to the Participant shall terminate immediately upon such violation, unless the Company otherwise determines. In addition, if, prior to the end of the term of an Option or Share Appreciation Right, the Participant is given notice by the Company or any of its Subsidiaries of the Participant's Termination of Service by the Company or any of its Subsidiaries for Cause, and the effective date of such Termination of Service is subsequent to the date of the delivery of such notice, the right of the Participant and the Participant's transferees to exercise any Option or Share Appreciation Right issued to the Participant shall be suspended from the time of the delivery of such notice until the earlier of (i) such time as it is determined or otherwise agreed that the Participant's service as a Service Provider will not be terminated for Cause as provided in such notice or (ii) the effective date of the Participant's Termination of Service by the Company or any of its Subsidiaries for Cause (in which case the right of the Participant and the Participant's transferees to exercise any Option or Share Appreciation Right issued to the Participant will terminate immediately upon the effective date of such termination of Service).

5.4 Exercise. Options and Share Appreciation Rights may be exercised by delivering to the Company a written notice of exercise, in a form the Administrator approves (which may be electronic), signed by the person authorized to exercise the Option or Share Appreciation Right, together with, as applicable, payment in full (i) as specified in Section 5.5 for the number of Shares for which the Award is exercised and (ii) as specified in Section 9.5 for any applicable taxes. Unless the Administrator otherwise determines, an Option or Share Appreciation Right may not be exercised for a fraction of a Share.

5.5 Payment Upon Exercise. Subject to Section 10.8, any Company insider trading policy (including blackout periods) and Applicable Laws, the exercise price of an Option must be paid by:

(a) cash, wire transfer of immediately available funds or by check payable to the order of the Company, provided that the Company may limit the use of one of the foregoing payment forms if one or more of the payment forms below is permitted;

(b) if there is a public market for Shares at the time of exercise, unless the Company otherwise determines, (A) delivery (including telephonically to the extent permitted by the Company) of an irrevocable and unconditional undertaking by a broker acceptable to the Company to deliver promptly to the Company sufficient funds to pay the exercise price, or (B) the Participant's delivery to the Company of a copy of irrevocable and unconditional instructions to a broker acceptable to the Company to deliver

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promptly to the Company cash or a check sufficient to pay the exercise price; provided that such amount is paid to the Company at such time as may be required by the Administrator;

(c) to the extent permitted by the Administrator, delivery (either by actual delivery or attestation) of Shares owned by the Participant valued at their Fair Market Value;

(d) to the extent permitted by the Administrator, surrendering Shares then issuable upon the Option's exercise valued at their Fair Market Value on the exercise date;

(e) to the extent permitted by the Administrator, delivery of a promissory note or any other property that the Administrator determines is good and valuable consideration; or

(f) to the extent permitted by the Company, any combination of the above payment forms approved by the Administrator.

ARTICLE VI.
RESTRICTED SHARES; RESTRICTED SHARE UNITS

6.1 General. The Administrator may grant Restricted Shares, or the right to purchase Restricted Shares, to any Service Provider, subject to the Company's right to repurchase all or part of such shares at their issue price or other stated or formula price from the Participant (or to require forfeiture of such shares) if conditions the Administrator specifies in the Award Agreement are not satisfied before the end of the applicable restriction period or periods that the Administrator establishes for such Award and subject to Applicable Laws. In addition, the Administrator may grant to Service Providers Restricted Share Units, which may be subject to vesting and forfeiture conditions during the applicable restriction period or periods, as set forth in an Award Agreement. The Administrator will determine and set forth in the Award Agreement the terms and conditions for each Restricted Share and Restricted Share Unit Award, subject to the conditions and limitations contained in the Plan.

6.2 Restricted Shares.

(a) Dividends. Participants holding shares of Restricted Shares will be entitled to all ordinary cash dividends paid with respect to such Shares, unless the Administrator provides otherwise in the Award Agreement. In addition, unless the Administrator provides otherwise, if any dividends or distributions are paid in Shares, or consist of a dividend or distribution to holders of Common Shares of property other than an ordinary cash dividend, the Shares or other property will be subject to the same restrictions on transferability and forfeitability as the shares of Restricted Shares with respect to which they were paid.

(b) Share Certificates. The Company may require that the Participant deposit in escrow with the Company (or its designee) any share certificates issued in respect of shares of Restricted Shares, together with a duly executed, but undated, instrument of transfer.

6.3 Restricted Share Units.

(a) Settlement. The Administrator may provide that settlement of Restricted Share Units will occur upon or as soon as reasonably practicable after the Restricted Share Units vest or will instead be deferred, on a mandatory basis or at the Participant's election, in a manner intended to comply with Section 409A.

(b) Shareholder Rights. A Participant will have no rights of a shareholder with respect to Shares subject to any Restricted Share Unit unless and until the Shares are delivered in settlement of the Restricted Share Unit.

(c) Dividend Equivalents. If the Administrator provides, a grant of Restricted Share Units may provide a Participant with the right to receive Dividend Equivalents. Dividend Equivalents may be paid currently or credited to an account for the Participant, settled in cash or Shares and subject to the same restrictions on transferability and forfeitability as the Restricted Share Units with respect to which the Dividend Equivalents are granted and subject to other terms and conditions as set forth in the Award Agreement.

ARTICLE VII.
OTHER SHARE OR CASH BASED AWARDS

Other Share or Cash Based Awards may be granted to Participants, including Awards entitling Participants to receive Shares to be delivered in the future and including annual or other periodic or long-term cash bonus awards (whether based on specified Performance Criteria or otherwise), in each case subject to any conditions and limitations in the Plan. Such Other Share or Cash Based Awards will also be available as a payment form in the settlement of other Awards, as standalone payments and as payment in lieu of compensation to which a Participant is otherwise entitled. Other Share or Cash Based Awards may be paid in Shares, cash or other property, as the Administrator determines. Subject to the provisions of the Plan, the Administrator will determine the terms and conditions of each Other Share or Cash Based Award, including any purchase price, performance goal (which may be based on the Performance Criteria), transfer restrictions, and vesting conditions, which will be set forth in the applicable Award Agreement.

ARTICLE VIII.
ADJUSTMENTS FOR CHANGES IN COMMON SHARES
AND CERTAIN OTHER EVENTS

8.1 Equity Restructuring. In connection with any Equity Restructuring, notwithstanding anything to the contrary in this Article VIII, the Administrator will equitably adjust each outstanding Award as it deems appropriate to reflect the Equity Restructuring, which may include adjusting the number and type of securities subject to each outstanding Award and/or the Award's exercise price or grant price (if applicable), granting new Awards to Participants, and making a cash payment to Participants. The adjustments provided under this Section 8.1 will be nondiscretionary and final and binding on the affected Participant and the Company; provided that the Administrator will determine whether an adjustment is equitable.

8.2 Corporate Transactions. In the event of any dividend or other distribution (whether in the form of cash, Common Shares, other securities, or other property), reorganization, merger, consolidation, combination, amalgamation, repurchase, recapitalization, liquidation, dissolution, or sale, transfer, exchange or other disposition of all or substantially all of the assets of the Company, or sale or exchange of Common Shares or other securities of the Company, Change in Control, issuance of warrants or other rights to purchase Common Shares or other securities of the Company, other similar corporate transaction or event, other unusual or nonrecurring transaction or event affecting the Company or its financial statements or any change in any Applicable Laws or accounting principles, the Administrator, on such terms and conditions as it deems appropriate, either by the terms of the Award or by action taken prior to the occurrence of such transaction or event (except that action to give effect to a change in Applicable Law or accounting principles may be made within a reasonable period of time after such change) and either automatically or upon the Participant's request, is hereby authorized to take any one or more of the following actions whenever the Administrator determines that such action is appropriate in order to (x)

prevent dilution or enlargement of the benefits or potential benefits intended by the Company to be made available under the Plan or with respect to any Award granted or issued under the Plan, (y) to facilitate such transaction or event or (z) give effect to such changes in Applicable Laws or accounting principles:

(a) To provide for the cancellation of any such Award in exchange for either an amount of cash or other property with a value equal to the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participant's rights under the vested portion of such Award, as applicable; provided that, if the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participant's rights, in any case, is equal to or less than zero, then the Award may be terminated without payment;

(b) To provide that such Award shall vest and, to the extent applicable, be exercisable as to all shares covered thereby, notwithstanding anything to the contrary in the Plan or the provisions of such Award;

(c) To provide that such Award be assumed by the successor or survivor corporation, or a parent or subsidiary thereof, or shall be substituted for by awards covering the shares of the successor or survivor corporation, or a parent or subsidiary thereof, with appropriate adjustments as to the number and kind of shares and/or applicable exercise or purchase price, in all cases, as determined by the Administrator;

(d) To make adjustments in the number and type of shares (or other securities or property) subject to outstanding Awards and/or with respect to which Awards may be granted under the Plan (including, but not limited to, adjustments of the limitations in Article IV hereof on the maximum number and kind of shares which may be issued) and/or in the terms and conditions of (including the grant or exercise price), and the criteria included in, outstanding Awards;

(e) To replace such Award with other rights or property selected by the Administrator; and/or

(f) To provide that the Award will terminate and cannot vest, be exercised or become payable after the applicable event.

8.3 Administrative Stand Still. In the event of any pending share dividend, bonus issue, share split, combination or exchange of shares, merger, amalgamation, consolidation or other distribution (other than normal cash dividends) of Company assets to shareholders, or any other extraordinary transaction or change affecting the Shares or the share price of Common Shares, including any Equity Restructuring or any securities offering or other similar transaction, for administrative convenience, the Administrator may refuse to permit the exercise of any Award for up to sixty days before or after such transaction.

8.4 General. Except as expressly provided in the Plan or the Administrator's action under the Plan, no Participant will have any rights due to any subdivision or consolidation of Shares of any class, dividend payment, increase or decrease in the number of Shares of any class or dissolution, liquidation, merger, amalgamation or consolidation of the Company or other corporation. Except as expressly provided with respect to an Equity Restructuring under Section 8.1 above or the Administrator's action under the Plan, no issuance by the Company of Shares of any class, or securities convertible into Shares of any class, will affect, and no adjustment will be made regarding, the number of Shares subject to an Award or the Award's grant or exercise price. The existence of the Plan, any Award Agreements and the Awards granted hereunder will not affect or restrict in any way the Company's right or power to make or authorize (i) any adjustment, recapitalization, reorganization or other change in the Company's capital structure or its business, (ii) any merger, amalgamation, consolidation dissolution or liquidation of the Company or sale of

Company assets or (iii) any sale or issuance of securities, including securities with rights superior to those of the Shares or securities convertible into or exchangeable for Shares. The Administrator may treat Participants and Awards (or portions thereof) differently under this Article VIII.

ARTICLE IX.
GENERAL PROVISIONS APPLICABLE TO AWARDS

9.1 Transferability. Except as the Administrator may determine or provide in an Award Agreement or otherwise for Awards other than Incentive Stock Options, Awards may not be sold, assigned, transferred, pledged or otherwise encumbered, either voluntarily or by operation of law, except by will or the laws of descent and distribution, or, subject to the Administrator's consent, pursuant to a domestic relations order, and, during the life of the Participant, will be exercisable only by the Participant. References to a Participant, to the extent relevant in the context, will include references to a Participant's authorized transferee that the Administrator specifically approves.

9.2 Documentation. Each Award will be evidenced in an Award Agreement, which may be written or electronic, as the Administrator determines. Each Award may contain terms and conditions in addition to those set forth in the Plan.

9.3 Discretion. Except as the Plan otherwise provides, each Award may be made alone or in addition or in relation to any other Award. The terms of each Award to a Participant need not be identical, and the Administrator need not treat Participants or Awards (or portions thereof) uniformly.

9.4 Termination of Status. The Administrator will determine how the disability, death, retirement, authorized leave of absence or any other change or purported change in a Participant's Service Provider status affects an Award and the extent to which, and the period during which, the Participant, the Participant's legal representative, conservator, guardian or Designated Beneficiary may exercise rights under the Award, if applicable.

9.5 Withholding. Each Participant must pay the Company, or make provision satisfactory to the Administrator for payment of, any taxes required by law to be withheld in connection with such Participant's Awards by the date of the event creating the tax liability. The Company may deduct a cash amount sufficient to satisfy such tax obligations based on the applicable statutory withholding rates (or such other rate as may be determined by the Company after considering any accounting consequences or costs) from any payment otherwise due to a Participant. Subject to Section 10.8 and any Company insider trading policy (including blackout periods), Participants may satisfy such tax obligations (i) in cash, by wire transfer of immediately available funds, by check made payable to the order of the Company, provided that the Company may limit the use of the foregoing payment forms if one or more of the payment forms below is permitted, (ii) to the extent permitted by the Administrator, in whole or in part by delivery of Shares valued at their Fair Market Value or elect to have the Company repurchase Shares otherwise issuable under an Award limited to the number of Common Shares which have a Fair Market Value on the date of repurchase necessary to pay the aggregate amount of tax liability, (iii) if there is a public market for Shares at the time the tax obligations are satisfied, unless the Company otherwise determines, (A) delivery (including telephonically to the extent permitted by the Company) of an irrevocable and unconditional undertaking by a broker acceptable to the Company to deliver promptly to the Company sufficient funds to satisfy the tax obligations, or (B) delivery by the Participant to the Company of a copy of irrevocable and unconditional instructions to a broker acceptable to the Company to deliver promptly to the Company cash or a check sufficient to satisfy the tax withholding; provided that such amount is paid to the Company at such time as may be required by the Administrator, or (iv) to the extent permitted by the Company, any combination of the foregoing payment forms approved by the Administrator. If any tax withholding obligation will be satisfied under clause (ii) of the immediately preceding sentence by the Company's retention of Shares

from the Award creating the tax obligation and there is a public market for Shares at the time the tax obligation is satisfied, the Company may elect to instruct any brokerage firm determined acceptable to the Company for such purpose to sell on the applicable Participant's behalf some or all of the Shares retained and to remit the proceeds of the sale to the Company or its designee, and each Participant's acceptance of an Award under the Plan will constitute the Participant's authorization to the Company and instruction and authorization to such brokerage firm to complete the transactions described in this sentence.

9.6 Amendment of Award; Repricing. The Administrator may amend, modify or terminate any outstanding Award, including by substituting another Award of the same or a different type, changing the exercise or settlement date, and converting an Incentive Stock Option to a Non-Qualified Share Option. The Participant's consent to such action will be required unless (i) the action, taking into account any related action, does not materially and adversely affect the Participant's rights under the Award, or (ii) the change is permitted under Article VIII or pursuant to Section 10.6. Notwithstanding the foregoing or anything in the Plan to the contrary, the Administrator may not except pursuant to Article VIII, without the approval of the shareholders of the Company, reduce the exercise price per share of outstanding Options or Share Appreciation Rights or cancel outstanding Options or Share Appreciation Rights in exchange for cash, other Awards or Options or Share Appreciation Rights with an exercise price per share that is less than the exercise price per share of the original Options or Share Appreciation Rights.

9.7 Conditions on Delivery of Shares. The Company will not be obligated to deliver any Shares under the Plan or remove restrictions from Shares previously delivered under the Plan until (i) all Award conditions have been met or removed to the Company's satisfaction, (ii) as determined by the Company, all other legal matters regarding the issuance and delivery of such Shares have been satisfied, including any applicable securities laws and stock exchange or stock market rules and regulations, and (iii) the Participant has executed and delivered to the Company such representations or agreements as the Administrator deems necessary or appropriate to satisfy any Applicable Laws. The Company's inability to obtain authority from any regulatory body having jurisdiction, which the Administrator determines is necessary to the lawful issuance and sale of any securities, will relieve the Company of any liability for failing to issue or sell such Shares as to which such requisite authority has not been obtained.

9.8 Acceleration. The Administrator may at any time provide that any Award will become immediately vested and fully or partially exercisable, free of some or all restrictions or conditions, or otherwise fully or partially realizable.

9.9 Additional Terms of Incentive Stock Options. The Administrator may grant Incentive Stock Options only to employees of the Company, any of its present or future parent or subsidiary corporations, as defined in Sections 424(e) or (f) of the Code, respectively, and any other entities the employees of which are eligible to receive Incentive Stock Options under the Code. If an Incentive Stock Option is granted to a Greater Than 10% Shareholder, the exercise price will not be less than 110% of the Fair Market Value on the Option's grant date, and the term of the Option will not exceed five years. All Incentive Stock Options will be subject to and construed consistently with Section 422 of the Code. By accepting an Incentive Stock Option, the Participant agrees to give prompt notice to the Company of dispositions or other transfers (other than in connection with a Change in Control) of Shares acquired under the Option made within (i) two years from the grant date of the Option or (ii) one year after the transfer of such Shares to the Participant, specifying the date of the disposition or other transfer and the amount the Participant realized, in cash, other property, assumption of indebtedness or other consideration, in such disposition or other transfer. Neither the Company nor the Administrator will be liable to a Participant, or any other party, if an Incentive Stock Option fails or ceases to qualify as an "incentive stock option" under Section 422 of the Code. Any Incentive Stock Option or portion thereof that fails to qualify as an "incentive stock option" under Section 422 of the Code for any reason, including becoming exercisable with respect

to Shares having a fair market value exceeding the $100,000 limitation under Treasury Regulation Section 1.422-4, will be a Non-Qualified Stock Option.

ARTICLE X.
MISCELLANEOUS

10.1 No Right to Employment or Other Status. No person will have any claim or right to be granted an Award, and the grant of an Award will not be construed as giving a Participant the right to continued employment or any other relationship with the Company. The Company expressly reserves the right at any time to dismiss or otherwise terminate its relationship with a Participant free from any liability or claim under the Plan or any Award, except as expressly provided in an Award Agreement.

10.2 No Rights as Shareholder; Certificates. Subject to the Award Agreement, no Participant or Designated Beneficiary will have any rights as a shareholder with respect to any Shares to be distributed under an Award until becoming the record holder of such Shares. Notwithstanding any other provision of the Plan, unless the Administrator otherwise determines or Applicable Laws require, the Company will not be required to deliver to any Participant certificates evidencing Shares issued in connection with any Award and instead such Shares may be recorded in the books of the Company (or, as applicable, its transfer agent or share plan administrator). The Company may place legends on share certificates issued under the Plan that the Administrator deems necessary or appropriate to comply with Applicable Laws.

10.3 Effective Date and Term of Plan. Unless earlier terminated by the Board, the Plan will become effective on the day prior to the Public Trading Date and will remain in effect until the tenth anniversary of the earlier of (i) the date the Board adopted the Plan or (ii) the date the Company's shareholders approved the Plan, but Awards previously granted may extend beyond that date in accordance with the Plan. If the Plan is not approved by the Company's shareholders, the Plan will not become effective, no Awards will be granted under the Plan and the Prior Plans will continue in full force and effect in accordance with their terms.

10.4 Amendment of Plan. The Administrator may amend, suspend or terminate the Plan at any time; provided that no amendment, other than an increase to the Overall Share Limit, may materially and adversely affect any Award outstanding at the time of such amendment without the affected Participant's consent. No Awards may be granted under the Plan during any suspension period or after Plan termination. Awards outstanding at the time of any Plan suspension or termination will continue to be governed by the Plan and the Award Agreement, as in effect before such suspension or termination. The Board will obtain shareholder approval of any Plan amendment to the extent necessary to comply with Applicable Laws.

10.5 Provisions for Non-U.S. Employees. The Administrator may modify Awards granted to Participants who are citizens or residents of a country other than the United States or employed outside the United States or establish subplans or procedures under the Plan to address differences in laws, rules, regulations or customs of such foreign jurisdictions with respect to tax, securities, currency, employee benefit or other matters.

10.6 Section 409A.

(a) General. The Company intends that all Awards be structured to comply with, or be exempt from, Section 409A, such that no adverse tax consequences, interest, or penalties under Section 409A apply. Notwithstanding anything in the Plan or any Award Agreement to the contrary, the Administrator may, without a Participant's consent, amend this Plan or Awards, adopt policies and procedures, or take any other actions (including amendments, policies, procedures and retroactive actions) as are necessary or appropriate to preserve the intended tax treatment of Awards, including any such actions

intended to (A) exempt this Plan or any Award from Section 409A, or (B) comply with Section 409A, including regulations, guidance, compliance programs and other interpretative authority that may be issued after an Award's grant date. The Company makes no representations or warranties as to an Award's tax treatment under Section 409A or otherwise. The Company will have no obligation under this Section 10.6 or otherwise to avoid the taxes, penalties or interest under Section 409A with respect to any Award and will have no liability to any Participant or any other person if any Award, compensation or other benefits under the Plan are determined to constitute noncompliant "nonqualified deferred compensation" subject to taxes, penalties or interest under Section 409A.

(b) Separation from Service. If an Award constitutes "nonqualified deferred compensation" under Section 409A, any payment or settlement of such Award upon a termination of a Participant's Service Provider relationship will, to the extent necessary to avoid taxes under Section 409A, be made only upon the Participant's "separation from service" (within the meaning of Section 409A), whether such "separation from service" occurs upon or after the termination of the Participant's Service Provider relationship. For purposes of this Plan or any Award Agreement relating to any such payments or benefits, references to a "termination," "termination of employment" or like terms means a "separation from service."

(c) Payments to Specified Employees. Notwithstanding any contrary provision in the Plan or any Award Agreement, any payment(s) of "nonqualified deferred compensation" required to be made under an Award to a "specified employee" (as defined under Section 409A and as the Administrator determines) due to his or her "separation from service" will, to the extent necessary to avoid taxes under Section 409A(a)(2)(B)(i) of the Code, be delayed for the six-month period immediately following such "separation from service" (or, if earlier, until the specified employee's death) and will instead be paid (as set forth in the Award Agreement) on the day immediately following such six-month period or as soon as administratively practicable thereafter (without interest). Any payments of "nonqualified deferred compensation" under such Award payable more than six months following the Participant's "separation from service" will be paid at the time or times the payments are otherwise scheduled to be made.

10.7 Limitations on Liability. Notwithstanding any other provisions of the Plan, no individual acting as a director, officer, other employee or agent of the Company or any Subsidiary will be liable to any Participant, former Participant, spouse, beneficiary, or any other person for any claim, loss, liability, or expense incurred in connection with the Plan or any Award, and such individual will not be personally liable with respect to the Plan because of any contract or other instrument executed in his or her capacity as an Administrator, director, officer, other employee or agent of the Company or any Subsidiary. The Company will indemnify and hold harmless each director, officer, other employee and agent of the Company or any Subsidiary that has been or will be granted or delegated any duty or power relating to the Plan's administration or interpretation, against any cost or expense (including attorneys' fees) or liability (including any sum paid in settlement of a claim with the Administrator's approval) arising from any act or omission concerning this Plan unless arising from such person's own fraud or bad faith.

10.8 Lock-Up Period. The Company may, at the request of any underwriter representative or otherwise, in connection with registering the offering of any Company securities under the Securities Act, prohibit Participants from, directly or indirectly, selling or otherwise transferring any Shares or other Company securities during a period of up to one hundred eighty days following the effective date of a Company registration statement filed under the

Securities Act, or such longer period as determined by the underwriter.

10.9 Data Privacy. As a condition for receiving any Award, each Participant explicitly and unambiguously consents to the collection, use and transfer, in electronic or other form, of personal data as described in this section by and among the Company and its Subsidiaries and affiliates exclusively for

implementing, administering and managing the Participant's participation in the Plan. The Company and its Subsidiaries and affiliates may hold certain personal information about a Participant, including the Participant's name, address and telephone number; birthdate; social security, insurance number or other identification number; salary; nationality; job title(s); any Shares held in the Company or its Subsidiaries and affiliates; and Award details, to implement, manage and administer the Plan and Awards (the "***Data***"). The Company and its Subsidiaries and affiliates may transfer the Data amongst themselves as necessary to implement, administer and manage a Participant's participation in the Plan, and the Company and its Subsidiaries and affiliates may transfer the Data to third parties assisting the Company with Plan implementation, administration and management. These recipients may be located in the Participant's country, or elsewhere, and the Participant's country may have different data privacy laws and protections than the recipients' country. By accepting an Award, each Participant authorizes such recipients to receive, possess, use, retain and transfer the Data, in electronic or other form, to implement, administer and manage the Participant's participation in the Plan, including any required Data transfer to a broker or other third party with whom the Company or the Participant may elect to deposit any Shares. The Data related to a Participant will be held only as long as necessary to implement, administer, and manage the Participant's participation in the Plan. A Participant may, at any time, view the Data that the Company holds regarding such Participant, request additional information about the storage and processing of the Data regarding such Participant, recommend any necessary corrections to the Data regarding the Participant or refuse or withdraw the consents in this Section 10.9 in writing, without cost, by contacting the local human resources representative. The Company may cancel Participant's ability to participate in the Plan and, in the Administrator's discretion, the Participant may forfeit any outstanding Awards if the Participant refuses or withdraws the consents in this Section 10.9. For more information on the consequences of refusing or withdrawing consent, Participants may contact their local human resources representative.

10.10 Severability. If any portion of the Plan or any action taken under it is held illegal or invalid for any reason, the illegality or invalidity will not affect the remaining parts of the Plan, and the Plan will be construed and enforced as if the illegal or invalid provisions had been excluded, and the illegal or invalid action will be null and void.

10.11 Governing Documents. If any contradiction occurs between the Plan and any Award Agreement or other written agreement between a Participant and the Company (or any Subsidiary) that the Administrator has approved, the Plan will govern, unless it is expressly specified in such Award Agreement or other written document that a specific provision of the Plan will not apply.

10.12 Governing Law. The Plan and all Awards will be governed by and interpreted in accordance with the laws of Delaware, disregarding any state's choice-of-law principles requiring the application of a jurisdiction's laws other than Delaware.

10.13 Claw-back Provisions. All Awards (including any proceeds, gains or other economic benefit the Participant actually or constructively receives upon receipt or exercise of any Award or the receipt or resale of any Shares underlying the Award) will be subject to any Company claw-back policy, including any claw-back policy adopted to comply with Applicable Laws (including the Dodd-Frank Wall Street Reform and Consumer Protection Act and any rules or regulations promulgated thereunder) as set forth in such claw-back policy or the Award Agreement.

10.14 Titles and Headings. The titles and headings in the Plan are for convenience of reference only and, if any conflict, the Plan's text, rather than such titles or headings, will control.

10.15 Conformity to Securities Laws. Participant acknowledges that the Plan is intended to conform to the extent necessary with Applicable Laws. Notwithstanding anything herein to the contrary, the Plan and all Awards will be administered only in conformance with Applicable Laws. To the extent

Applicable Laws permit, the Plan and all Award Agreements will be deemed amended as necessary to conform to Applicable Laws.

10.16 Relationship to Other Benefits. No payment under the Plan will be taken into account in determining any benefits under any pension, retirement, savings, profit sharing, group insurance, welfare or other benefit plan of the Company or any Subsidiary except as expressly provided in writing in such other plan or an agreement thereunder.

10.17 Broker-Assisted Sales. In the event of a broker-assisted sale of Shares in connection with the payment of amounts owed by a Participant under or with respect to the Plan or Awards, including amounts to be paid under the final sentence of Section 9.5: (a) any Shares to be sold through the broker-assisted sale will be sold on the day the payment first becomes due, or as soon thereafter as practicable; (b) such Shares may be sold as part of a block trade with other Participants in the Plan in which all participants receive an average price; (c) the applicable Participant will be responsible for all broker's fees and other costs of sale, and by accepting an Award, each Participant agrees to indemnify and hold the Company harmless from any losses, costs, damages, or expenses relating to any such sale; (d) to the extent the Company or its designee receives proceeds of such sale that exceed the amount owed, the Company will pay such excess in cash to the applicable Participant as soon as reasonably practicable; (e) the Company and its designees are under no obligation to arrange for such sale at any particular price; and (f) in the event the proceeds of such sale are insufficient to satisfy the Participant's applicable obligation, the Participant may be required to pay immediately upon demand to the Company or its designee an amount in cash sufficient to satisfy any remaining portion of the Participant's obligation.

ARTICLE XI.
DEFINITIONS

As used in the Plan, the following words and phrases will have the following meanings:

11.1 "***Administrator***" means the Board or a Committee to the extent that the Board's powers or authority under the Plan have been delegated to such Committee.

11.2 "***Applicable Laws***" means the requirements relating to the administration of equity incentive plans under U.S. federal and state securities, tax and other applicable laws, rules and regulations, the applicable rules of any stock exchange or quotation system on which the Common Shares are listed or quoted and the applicable laws and rules of any foreign country or other jurisdiction where Awards are granted or issued under the Plan, including without limitation, the laws of Bermuda.

11.3 "***Award***" means, individually or collectively, a grant under the Plan of Options, Share Appreciation Rights, Restricted Shares, Restricted Share Units or Other Share or Cash Based Awards.

11.4 "***Award Agreement***" means a written agreement evidencing an Award, which may be electronic, that contains such terms and conditions as the Administrator determines, consistent with and subject to the terms and conditions of the Plan.

11.5 "***Board***" means the Board of Directors of the Company.

11.6 "***Cause***" means, with respect to a Participant, (A) dishonesty with respect to the Company or any Subsidiary; (B) insubordination, substantial malfeasance or nonfeasance of duty; (C) unauthorized disclosure of confidential information; (D) breach by a Participant of any provision of any employment, consulting, advisory, nondisclosure, non-competition or similar agreement between the Participant and the Company or any Subsidiary; and (E) conduct substantially prejudicial to the business of the Company or

any Subsidiary; provided, however, that any provision in an agreement between a Participant and the Company or an Subsidiary, which contains a conflicting definition of Cause for termination and which is in effect at the time of such termination, shall supersede this definition with respect to that Participant. The determination of the Administrator as to the existence of Cause will be conclusive on the Participant and the Company.

11.7 "***Change in Control***" means (a) a sale of all or substantially all of the Company's assets, or (b) any merger, amalgamation, consolidation or other business combination transaction of the Company with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the voting shares of the Company outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into voting shares of the surviving entity) a majority of the total voting power represented by the voting shares of the Company (or the surviving entity) outstanding immediately after such transaction, or (c) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of the Company. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur: (A) on account of the acquisition of voting shares by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Company's voting shares in a transaction or series of related transactions that are primarily a private financing transaction for the Company or (B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "***Subject Person***"), exceeds the designated percentage threshold of the outstanding voting shares as a result of a repurchase or other acquisition of voting shares by the Company reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operating of this sentence) as a result of the acquisition of voting shares by the Company, and after such share acquisition, the Subject Person becomes the owner of any additional voting shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

Notwithstanding the foregoing, if a Change in Control constitutes a payment event with respect to any Award (or portion of any Award) that provides for the deferral of compensation that is subject to Section 409A, to the extent required to avoid the imposition of additional taxes under Section 409A, the transaction or event described in subsection (a), (b) or (c) with respect to such Award (or portion thereof) shall only constitute a Change in Control for purposes of the payment timing of such Award if such transaction also constitutes a "change in control event," as defined in Treasury Regulation Section 1.409A-3(i)(5).

The Administrator shall have full and final authority, which shall be exercised in its discretion, to determine conclusively whether a Change in Control has occurred pursuant to the above definition, the date of the occurrence of such Change in Control and any incidental matters relating thereto; provided that any exercise of authority in conjunction with a determination of whether a Change in Control is a "change in control event" as defined in Treasury Regulation Section 1.409A-3(i)(5) shall be consistent with such regulation.

11.8 "***Code***" means the Internal Revenue Code of 1986, as amended, and the regulations issued thereunder.

11.9 "***Committee***" means one or more committees or subcommittees of the Board, which may include one or more Company directors or executive officers, to the extent Applicable Laws permit. To the extent required to comply with the provisions of Rule 16b-3, it is intended that each member of the Committee will be, at the time the Committee takes any action with respect to an Award that is subject to

Rule 16b-3, a "non-employee director" within the meaning of Rule 16b-3; however, a Committee member's failure to qualify as a "non-employee director" within the meaning of Rule 16b-3 will not invalidate any Award granted by the Committee that is otherwise validly granted under the Plan.

11.10 "***Common Shares***" means the Class A Common Shares of the Company of par value US$0.0001 each.

11.11 "***Company***" means Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company, or any successor.

11.12 "***Consultant***" means any person, including any adviser, engaged by the Company or its parent or Subsidiary to render services to such entity if the consultant or adviser: (i) renders bona fide services to the Company; (ii) renders services not in connection with the offer or sale of securities in a capital-raising transaction and does not directly or indirectly promote or maintain a market for the Company's securities; and (iii) is a natural person.

11.13 "***Designated Beneficiary***" means the beneficiary or beneficiaries the Participant designates, in a manner the Administrator determines, to receive amounts due or exercise the Participant's rights if the Participant dies or becomes incapacitated. Without a Participant's effective designation, "Designated Beneficiary" will mean the Participant's estate.

11.14 "***Director***" means a Board member.

11.15 "***Disability***" means a permanent and total disability under Section 22(e)(3) of the Code, as amended.

11.16 "***Dividend Equivalents***" means a right granted to a Participant under the Plan to receive the equivalent value (in cash or Shares) of dividends paid on Shares.

11.17 "***Employee***" means any employee of the Company or its Subsidiaries.

11.18 "***Equity Restructuring***" means a nonreciprocal transaction between the Company and its shareholders, such as a share dividend, bonus issue, share split, spin-off or recapitalization through a large, nonrecurring cash dividend, that affects the number or kind of Shares (or other Company securities) or the share price of Common Shares (or other Company securities) and causes a change in the per share value of the Common Shares underlying outstanding Awards.

11.19 "***Exchange Act***" means the Securities Exchange Act of 1934, as amended.

11.20 "***Fair Market Value***" means, as of any date, the value of Common Shares determined as follows: (i) if the Common Shares are listed on any established stock exchange, a share's Fair Market Value will be the closing sales price for such Common Shares as quoted on such exchange for such date, or if no sale occurred on such date, the last day preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; (ii) if the Common Shares are not traded on a stock exchange but is quoted on a national market or other quotation system, the closing sales price on such date, or if no sales occurred on such date, then on the last date preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; or (iii) without an established market for the Common Shares, the Administrator will determine the Fair Market Value in its discretion. Notwithstanding the foregoing, with respect to any Award granted on the pricing date of the Company's initial public offering, the Fair Market Value shall

mean the initial public offering price of a Share as set forth in the Company's final prospectus relating to its initial public offering filed with the Securities and Exchange Commission.

11.21 "***Greater Than 10% Shareholder***" means an individual then owning (within the meaning of Section 424(d) of the Code) more than 10% of the total combined voting power of all classes of shares of the Company or its parent or subsidiary corporation, as defined in Section 424(e) and (f) of the Code, respectively.

11.22 "***Incentive Stock Option***" means an Option intended to qualify as an "incentive stock option" as defined in Section 422 of the Code.

11.23 "***Non-Qualified Stock Option***" means an Option not intended or not qualifying as an Incentive Stock Option.

11.24 "***Option***" means an option to purchase Shares.

11.25 "***Other Share or Cash Based Awards***" means cash awards, awards of Shares, and other awards valued wholly or partially by referring to, or are otherwise based on, Shares or other property.

11.26 "***Overall Share Limit***" means (i) 4,446,530 Common Shares, (ii) any Common Shares which are subject to Prior Plan Awards which become available for issuance under the Plan pursuant to Article IV and (iii) an annual increase on the first day of each calendar year beginning January 1, 2019 and ending on and including January 1, 2028, equal to the lesser of (A) 4% of the aggregate number of Shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (B) such smaller number of Shares as is determined by the Board.

11.27 "***Participant***" means a Service Provider who has been granted an Award.

11.28 "***Performance Criteria***" mean the criteria (and adjustments) that the Administrator may select for an Award to establish performance goals for a performance period, which may include the following: net earnings or losses (either before or after one or more of interest, taxes, depreciation, amortization, and non-cash equity-based compensation expense); gross or net sales or revenue or sales or revenue growth; net income (either before or after taxes) or adjusted net income; profits (including but not limited to gross profits, net profits, profit growth, net operation profit or economic profit), profit return ratios or operating margin; budget or operating earnings (either before or after taxes or before or after allocation of corporate overhead and bonus); cash flow (including operating cash flow and free cash flow or cash flow return on capital); return on assets; return on capital or invested capital; cost of capital; return on shareholders' equity; total shareholder return; return on sales; costs, reductions in costs and cost control measures; expenses; working capital; earnings or loss per share; adjusted earnings or loss per share; price per share or dividends per share (or appreciation in or maintenance of such price or dividends); regulatory achievements or compliance; implementation, completion or attainment of objectives relating to research, development, regulatory, commercial, or strategic milestones or developments; market share; economic value or economic value added models; division, group or corporate financial goals; customer satisfaction/growth; customer service; employee satisfaction; recruitment and maintenance of personnel; human resources management; supervision of litigation and other legal matters; mergers, acquisitions, and other strategic partnerships, licenses, and other transactions; financial ratios (including those measuring liquidity, activity, profitability or leverage); debt levels or reductions; sales-related goals; financing and other capital raising transactions; cash on hand; acquisition activity; investment sourcing activity; and marketing initiatives, any of which may be measured in absolute terms or as compared to any incremental increase or decrease. Such performance goals also may be based solely by reference to the Company's performance or the performance of a Subsidiary, division, business segment or business unit of the

Company or a Subsidiary, or based upon performance relative to performance of other companies or upon comparisons of any of the indicators of performance relative to performance of other companies. The Committee may provide for exclusion of the impact of an event or occurrence which the Committee determines should appropriately be excluded, including (a) restructurings, discontinued operations, extraordinary items, and other unusual, infrequently occurring or non-recurring charges or events, (b) asset write-downs, (c) litigation or claim judgments or settlements, (d) acquisitions or divestitures, (e) reorganization or change in the corporate structure or capital structure of the Company, (f) an event either not directly related to the operations of the Company, Subsidiary, division, business segment or business unit or not within the reasonable control of management, (g) foreign exchange gains and losses, (h) a change in the fiscal year of the Company, (i) the refinancing or repurchase of bank loans or debt securities, (j) unbudgeted capital expenditures, (k) the issuance or repurchase of equity securities and other changes in the number of issued and outstanding shares, (l) conversion of some or all of convertible securities to Common Shares, (m) any business interruption event (n) the cumulative effects of tax or accounting changes in accordance with U.S. generally accepted accounting principles, or (o) the effect of changes in other laws or regulatory rules affecting reported results.

11.29 "***Plan***" means this 2018 Incentive Award Plan.

11.30 "***Prior Plans***" means, collectively, the Kiniksa Pharmaceuticals, Ltd. 2015 Equity Incentive Plan and any prior equity incentive plans of the Company or its predecessor.

11.31 "***Prior Plan Award***" means an award outstanding under the Prior Plans as of the Plan's effective date in Section 10.3.

11.32 "***Public Trading Date***" means the first date upon which the Common Shares are listed (or approved for listing) upon notice of issuance on any securities exchange or designated (or approved for designation) upon notice of issuance as a national market security on an interdealer quotation system, or, if earlier, the date on which the Company becomes a "publicly held corporation" for purposes of Treasury Regulation Section 1.162-27(c)(1).

11.33 "***Restricted Share***" means a Share awarded to a Participant under Article VI subject to certain vesting conditions and other restrictions.

11.34 "***Restricted Share Unit***" means an unfunded, unsecured right to receive, on the applicable settlement date, one Share or an amount in cash or other consideration determined by the Administrator to be of equal value as of such settlement date, subject to certain vesting conditions and other restrictions.

11.35 "***Rule 16b-3***" means Rule 16b-3 promulgated under the Exchange Act.

11.36 "***Section 409A***" means Section 409A of the Code and all regulations, guidance, compliance programs and other interpretative authority thereunder.

11.37 "***Securities Act***" means the Securities Act of 1933, as amended.

11.38 "***Service Provider***" means an Employee, Consultant or Director.

11.39 "***Shares***" means Common Shares.

11.40 "***Share Appreciation Right***" means a share appreciation right granted under Article V.

11.41 "***Subject Person***" has the meaning set forth in Section 11.7.

11.42 "***Subsidiary***" means any entity (other than the Company), whether domestic or foreign, in an unbroken chain of entities beginning with the Company if each of the entities other than the last entity in the unbroken chain beneficially owns, at the time of the determination, securities or interests representing at least 50% of the total combined voting power of all classes of securities or interests in one of the other entities in such chain.

11.43 "***Substitute Awards***" shall mean Awards granted or Shares issued by the Company in assumption of, or in substitution or exchange for, awards previously granted, or the right or obligation to make future awards, in each case by a company acquired by the Company or any Subsidiary or with which the Company or any Subsidiary combines.

11.44 "***Termination of Service***" means the date the Participant ceases to be a Service Provider.

* * * * *

Exhibit 10.3

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of May , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Sanj K. Patel (the "Executive").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Executive possesses certain experience and expertise that qualify him to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Executive are party to an Amended and Restated Employment Agreement dated as of December June 29, 2017 (the "Original Agreement"); and

WHEREAS, the Company and the Executive wish to amend and restate the Original Agreement in accordance with the terms set forth herein and the Executive shall continue to serve as its Chief Executive Officer on the terms and conditions set forth in this Agreement, and the Executive wishes to continue such employment.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the parties hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Executive's gross negligence or willful misconduct in performance of his duties to the Company, where such gross negligence or willful misconduct has resulted in material damage to the Company or any of its Affiliates or successors; or

(ii) The Executive's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Executive's commission of a felony or crime involving moral turpitude; or

(iv) The Executive's material breach of any provision of this Agreement or any other written agreement between the Executive and the Company; or

(v) The Executive's failure to comply with lawful directives of the Parent Board (as defined below), which has caused damage to the Company or any of its Affiliates or successors.

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets, or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction;

(iii) A change in the composition of the Parent Board such that the individuals who, as of the Effective Date, constitute the Parent Board (such Parent Board shall be hereinafter referred to as the "Incumbent Board") cease for any reason to constitute at least a majority of the Parent Board; provided, however, that, for purposes of this Agreement, any individual who becomes a member of the Parent Board subsequent to the Effective Date, whose election, or nomination for election by the Parent's shareholders, was approved by a vote of at least a majority of those individuals who are members of the Incumbent Board (or deemed to be such pursuant to this proviso) shall be considered as though such individual were a member of the Incumbent Board; or

(iv) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operating of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of ERISA, as amended from time-to-time.

(f) "Founder Invention and Non-Disclosure Agreement" shall mean the Founder Invention and Non-Disclosure Agreement, dated as of September 16, 2015.

(g) "Founder Non-Competition and Non-Solicitation Agreement" shall mean the Founder Non-Competition and Non-Solicitation Agreement, dated as of September 16, 2015.

(h) "Good Reason" shall mean any of the following, occurring without the Executive's written consent:

(i) the assignment to the Executive of duties materially inconsistent with the Executive's title, position, status, reporting relationships, authority, duties or responsibilities as contemplated by Section 3, or any other action by the Company which results in a diminution in the Executive's title, position, status, reporting relationships, authority, duties or responsibilities; or

(ii) a requirement that the Executive relocate his primary reporting location to a location more than fifty (50) miles from the location of the Company's offices in Lexington, Massachusetts as of the Effective Date; or

(iii) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b) 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith which are remedied by the Company promptly after receipt of notice thereof given by the Executive; or

(iv) a material diminution in the budget over which Executive has responsibility; or

(v) a breach by the Company of this Agreement between the Company and Executive.

(i) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(j) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Executive hereby accepts employment subject to the terms and conditions set forth in this Agreement. Executive's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Executive shall serve the Company as its Chief Executive Officer and shall report to the Board of Directors of the Parent (the "Parent Board"). During the Term, the Executive shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of his position.

(b) The Executive is currently in the role of Chairman of the Parent Board and the Company, and shall continue to serve as a member of the Parent Board and the Company. The Parent shall propose to the shareholders of the Parent Board at each appropriate annual meeting of such shareholders during Executive's employment the reelection of the Executive as a director of the Parent Board, provided that the Executive is otherwise eligible for such election.

(c) In addition, and without further compensation, the Executive shall serve as a director and/or officer of one or more of the Company's Affiliates if so elected or appointed from time-to-time.

(d) During the Term, the Executive shall devote his full business time and his best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of his duties and responsibilities hereunder. The Executive shall not engage in any other business activity or serve in any industry, trade, professional, governmental or academic position during the term of this Agreement, except such activities as shall not interfere with the performance of his duties to the Company. Notwithstanding the foregoing, the Executive shall be entitled to attend to personal and family affairs and investments, be involved in not-for-profit, charitable and professional activities and serve on up to two (2) for-profit boards, provided that the foregoing does not, individually or in the aggregate, materially interfere with Executive's responsibilities under this Agreement.

(e) Immediately upon termination of Executive's employment with the Company for any reason, Executive will be deemed to resign any and all positions held by him, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Executive during the Term and subject to the Executive's performance of his duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Executive with the following compensation and benefits:

(a) Base Salary. During the Term, the Company shall pay the Executive at the rate of not less than $750,000 per annum, payable in accordance with the payroll practices of the Company for its executives and subject to increase from time-to-time by the Parent Board, in its sole discretion (such base salary, as from time-to-time increased, the "Base Salary"). The Base Salary shall not be reduced at any time (including after any such increase) without the Executive's prior written consent, and the term Base Salary as used in this Agreement shall refer to Executive's Base Salary as so increased.

(b) Bonus Compensation. During the Employment Period, the Executive shall be paid an annual cash bonus ("Annual Bonus") with a minimum target level of 60% of Annual Base Salary (the "Target Bonus"). The applicable corporate and individual performance targets shall be determined by the Compensation Committee of the Parent Board (the "Compensation Committee") after consultation with the Executive, within the first ninety (90) days of each calendar year. The actual Annual Bonus for each calendar year shall be determined in good faith by the Compensation Committee based upon actual corporate and individual performance against established objective for such year (and, if determined by the Compensation Committee, may exceed the Target Bonus) and shall be payable in accordance with the procedures specified by the Compensation Committee; provided that the Annual Bonus shall be paid to Executive no later than March 15th of the calendar year immediately following the calendar year in which it was earned.

(c) Equity Participation. Executive shall be eligible to receive equity awards annually under the Parent's 2018 Incentive Award Plan or any other equity plan (collectively with the Parent's 2015 Equity Incentive Plan, the "Equity Plan"), and the award agreements related to such plans, as determined in the discretion of the Parent Board, commencing upon the Effective Date. Notwithstanding the provisions of the Equity Plan or any agreement or award, Executive shall be immediately 100% fully vested in all equity awards granted to him under this Agreement or any other agreement that vest solely based on the passage of time in the event of a Change in Control in which the

award is not being assumed or substituted as provided for in the applicable Equity Plan (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(d) Vacation. During the Term, the Executive shall be entitled to earn vacation at the rate of five (5) weeks per year, to be taken at such times and intervals as shall be determined by the Executive, subject to the reasonable business needs of the Company. The Executive shall be entitled to carryover up to two (2) weeks of vacation into the following calendar year. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(e) Other Benefits. During the Term, the Executive shall be entitled to participate in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Executive (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or delete its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Executive.

(f) Business Expenses. The Company shall promptly pay or reimburse the Executive for all reasonable business expenses incurred or paid by the Executive in the performance of his duties and responsibilities hereunder, subject to reasonable substantiation and documentation, as may be specified by the Company from time-to-time.

5. Termination of Employment and Severance Benefits. The Executive's employment hereunder shall terminate under the following circumstances:

(a) Death. In the event of the Executive's death, the Executive's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Executive's employment hereunder, upon notice to the Executive, in the event that the Executive becomes disabled during his employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of his duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Executive's place during any period of the Executive's disability. Notwithstanding any such designation, the Executive shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(e), to the extent permitted by the then-current terms of the applicable benefit plans, until the Executive becomes eligible for disability income benefits under the Company's disability income plan or until the termination of his employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan of the Company, the Executive shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(e) and the terms of such plans, until the termination of his employment.

(c) By the Company for Cause. The Company may terminate the Executive's employment hereunder for Cause at any time upon notice to the Executive setting forth in reasonable detail

the nature of such Cause. In the cases of Section 1(b)(i), 1(b)(iv), or 1(b)(v) above, the Company may not effectuate the termination for Cause unless and until: (i) the Company provides Executive with written notice setting forth in reasonable detail the nature for such Cause which must be given to Executive no later than the thirtieth (30) day following the initial commission of the acts or omissions constituting Cause, and (ii) Executive fails to cure the alleged conduct or events supporting Cause within thirty (30) days following Executive's receipt of such notice.

(d) By the Company Other than for Cause. The Company may terminate the Executive's employment hereunder other than for Cause at any time upon written notice to the Executive.

(e) By the Executive for Good Reason. The Executive may terminate his employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating his employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

(f) By the Executive Other than for Good Reason. The Executive may terminate his employment hereunder at any time upon sixty (60) days' notice to the Company. In the event of termination of the Executive pursuant to this Section 5(f), the Parent Board or the Company may elect to waive the period of notice, or any portion thereof. In the event that the Parent Board or the Company so waives some or all of the period of notice, the Company shall pay the Executive his Base Salary for the period so waived.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Executive's employment for any reason, the Company shall pay to the Executive: (i) any Base Salary earned but not paid during the final payroll period of the Executive's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Annual Bonus due to Executive for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Executive but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Executive at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any business expenses due under this Section 6(a) shall be paid within sixty (60) days following the date of termination. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Executive hereunder.

(b) Severance. In the event the Executive's employment terminates pursuant to Section 5(a), 5(b), 5(d) or 5(e) of this Agreement, in addition to Final Compensation, the Company (i) shall accelerate the vesting of all unvested equity awards that vest solely based on the passage of time either then held by, or previously granted to, Executive (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards) by eighteen (18) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) shall pay the Executive (A) a lump sum equal to the Base Salary plus the Target Bonus divided by twelve (12), then multiplied by the number of months set forth in the Severance Period (as defined below) (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below) and (C) an additional one-time bonus of $25,000 (the "One-Time Bonus"). Subject to Sections 6(e) and 7(a) of this Agreement (i) the Severance Payment and the One-Time Bonus shall be paid on the sixtieth (60th) day following the date of termination

and (ii) the Post-Termination Bonus shall be paid at the time provided in the applicable Bonus Plan or form of annual award issued thereunder, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Equity Plan, the Executive shall be immediately 100% fully vested in all unvested equity awards that vest solely based on the passage of time of Parent or its Affiliates (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Executive either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(c) Severance Period. For the purposes of this Agreement, the "Severance Period" shall be twenty-four (24) months.

(d) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the year in which the termination occurs.

(e) Release of Claims. Executive's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Executive's signing and returning to the Company a general release of claims in the form provided by the Company at the time the Executive's employment is terminated (the "Employee Release"). The Executive must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein, which shall be no longer than seven (7) days from the date of the Executive's signature

(f) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Executive's employment hereunder. Except for any right of the Executive to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Executive's employment without regard to the payment of any Severance Payment or Post-Termination Bonus.

(g) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Executive under Sections 8 hereof. The obligation of the Company to make, and the right of the Executive to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Executive's continued full performance of obligations under Section 8, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Executive's termination of employment, the Executive is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Executive's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Executive's employment with the Company, the Executive has executed the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement. The parties hereto acknowledge and agree that this Agreement, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement shall be considered separate contracts. In addition, Executive represents and warrants that he shall be able to and will perform the duties of this position without utilizing any material confidential and/or proprietary information that Executive may have obtained in connection with employment with any prior employer, and that he shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Executive's employment, Executive shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Executive, provided, that the Executive will not have an obligation under this paragraph with respect to any claim in which the Executive has filed directly against the Company or related persons or entities. The Executive's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Executive's employment, Executive also shall reasonably

cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Executive was employed by the Company, provided Executive will not have any obligation under this paragraph with respect to any claim in which Executive has filed directly against the Company or related persons or entities. The Company shall reimburse Executive for any reasonable out-of-pocket expenses incurred in connection with Executive's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Executive shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Executive's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Executive. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Executive as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Executive and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Executive and the Company within fifteen (15) calendar days after the date on which Executive's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Executive or the Company) or such other time as requested by Executive or the Company.

(d) If Executive receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Executive agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Executive shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Executive to the extent provided in its then current Certificate of Incorporation or By-Laws. The Executive agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of his employment with the Company.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Executive may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Executive in the event that the Executive is transferred to a position with any of the Affiliates or in the event that the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Executive, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a

reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Executive at his last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement, constitutes the entire understanding and agreement of the parties hereto regarding the employment of Executive. This Agreement supersedes all prior negotiations, discussions, correspondence, communications, understandings, and agreements between the parties (including any offer letter given to Executive) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Executive acknowledge that the Executive holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Executive under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Executive and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof.

[Signature page follows immediately.]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Executive, as of the date first above written.

EXECUTIVE

Sanj K. Patel

KINIKSA PHARMACEUTICALS CORP.

By: _____

Name: Thomas W. Beetham
Title: Executive Vice President, Chief Legal Officer and Corporate Development

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of May , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Stephen F. Mahoney (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee is currently serving as the Company's President and Chief Operating Officer and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the "Original Agreement"); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

EVP (Founder)

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Founder Invention and Non-Disclosure Agreement" shall mean the Founder Invention and Non-Disclosure Agreement between the Employee and Company dated as of September 18, 2015.

(g) "Founder Non-Competition and Non-Solicitation Agreement" shall mean the Founder Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of September 18, 2015.

(h) "Good Reason" shall mean any of the following, occurring without the Employee's written consent:

(i) a demotion of the Employee to a position with responsibilities substantially less than the Employee's prior position; provided that, a change in title, reporting relationships and/or responsibilities of the Employee could, but do not necessarily in and of themselves, individually or in the aggregate, constitute a demotion for purposes of this Section 1(h)(i); and in all instances, the determination of whether a demotion has occurred shall be made by the Company in good faith; or

(ii) a requirement that the Employee relocate Employee's primary reporting location to a location more than fifty (50) miles from the location of the Company's current offices in Lexington, Massachusetts as of the Effective Date; or

(iii) a reduction of more than five percent (5%) of Employee's Base Salary, other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated to Employee; or

(iv) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b), 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith that are remedied by the Company promptly after receipt of notice thereof given by the Employee; or

(v) a breach by the Company of this Agreement.

(i) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(j) "Parent Board" shall mean the board of directors of the Parent.

(k) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its President and Chief Operating Officer, and shall initially report to the Chief Executive Officer of the Company. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business

activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $442,260, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with an initial target level of 40% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee for Good Reason. The Employee may terminate Employee's employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating Employee's employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

(f) By the Employee Other than for Good Reason. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(f), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b), 5(d) or 5(e) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below) and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Founder Invention and Non-Disclosure Agreement and the Founder Invention and Non-Disclosure Agreement, both of which the Company and Employee acknowledge and agree shall be considered separate contracts. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the

reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company. The parties acknowledge that the Employee is also indemnified by the Parent to the extent set forth in the Indemnification Agreement between the Parent and Employee dated December 16, 2015.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement and the Founder Non-Solicitation and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior

negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that the Employee holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE

Name: Stephen F. Mahoney

KINIKSA PHARMACEUTICALS CORP.

By:

Name: Sanj K. Patel
Title: Chief Executive Officer

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of May , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and John F. Paolini (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee is currently serving as the Company's Senior Vice President and Chief Medical Officer and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the "Original Agreement"); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

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(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Confidential Information and Non-Competition Agreement" shall mean the Employee Proprietary Information, Inventions Assignment, Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of August 15, 2016.

(g) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(h) "Parent Board" shall mean the board of directors of the Parent.

(i) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its Senior Vice President and Chief Medical Officer, and shall initially report to the Chief Operating Officer of the Company. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $420,000, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with a target level of 30% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year immediately

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following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(e), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b) or 5(d) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary divided by twelve (12), then multiplied by the number of months of the Severance Period (as defined below) (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below), and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the

Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement). The "Severance Period" shall be nine (9) months; provided, that if the Employee's separation from service occurs during the twelve (12) months following a Change in Control, then the Severance Period shall be twelve (12) months.

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8 and the Confidential Information and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months

following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Confidential Information and Non-Competition Agreement, which the Company and Employee acknowledge and agree shall be considered a separate contract. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited

to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall

reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not

prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Confidential Information and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that options and other equity awards have been and may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE

Name: John F. Paolini

KINIKSA PHARMACEUTICALS CORP.

By: _____

Name: Sanj K. Patel

CONFIDENTIAL

ASSET PURCHASE AGREEMENT

between

KINIKSA PHARMACEUTICALS, LTD.

and

BIOGEN MA INC.

dated as of September 7, 2016

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

TABLE OF CONTENTS

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULES

EXHIBITS

ASSET PURCHASE AGREEMENT

This Asset Purchase Agreement (this "***Agreement***") is made and entered into as of September 7, 2016 (the "***Effective Date***"), between Biogen MA Inc., a Massachusetts corporation ("***Biogen***"), and Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company ("***Kiniksa***"). Kiniksa and Biogen are sometimes referred to herein individually as a "***Party***" and collectively as the "***Parties***".

RECITALS

WHEREAS, Biogen is engaged, among other things, in the development of BIIB069 (as defined below); and

WHEREAS, Biogen desires to sell to Kiniksa, and Kiniksa desires to purchase from Biogen, certain assets of Biogen used in or relating to BIIB069 and Kiniksa is willing to assume certain liabilities of Biogen relating to BIIB069, all upon the terms and conditions set forth herein.

NOW, THEREFORE, in consideration of the premises and the mutual covenants, representations and warranties herein contained, and for other good and valuable consideration, the receipt and sufficiency of which are hereby mutually acknowledged, the Parties hereby agree as follows:

1. DEFINITIONS

1.1 **Defined Terms**. As used in this Agreement, the following defined terms shall have the meanings specified below:

"***Acquired Antibody***" means (a) BIIB069 and [***] Antibody that is Covered by one or more claims within the Acquired Patent Rights and (b) [***] BIIB069 or [***] that, in each case, [***].

"***Acquired Know-How***" means all Know-How that is (a) owned or Controlled by Biogen as of the Effective Date, and (b) used or generated by Biogen solely in connection with its research and development of BIIB069 prior to the Effective Date. The Acquired Know-How is described on Part 2 of Schedule A attached hereto.

"***Acquired Patent Rights***" means (a) the patent applications listed on Part 1 of Schedule A attached hereto and (b) any divisionals, continuations, continuations-in-part, substitutions, patents of addition, reissues, extensions, re-examinations or renewal applications related to, or claiming priority to, the foregoing (including any supplemental patent certificates) or any confirmation patent or registration patent, and all patents issuing on, and all foreign counterparts of, any of the foregoing.

"***Affiliate***" means, with respect to any Person, any other Person which controls, is controlled by or is under common control with such Person, for as long as such control exists.

For purposes of this definition, "control" shall mean the direct or indirect ownership of more than fifty percent (50%) of the voting or economic interest of a Person, or the power, whether pursuant to contract, ownership of securities or otherwise, to direct the management and policies of a Person. For clarity, once a Person ceases to be an Affiliate of a Party, then, without any further action, such Person shall cease to have any rights under this Agreement by reason of being an Affiliate of such Party**.**

"***Annual Net Sales***" shall mean the cumulative worldwide Net Sales of an applicable Product in any Calendar Year.

"***Antibody***" means any immunoglobulin molecule [***] whether in [***] or any other [***] form, and will include (a) any [***] (b) any [***], and (c) any [***].

"***Applicable Multiplier***" means the percentage used to determine the portion of the milestone payments and royalty payments due and payable by Biogen with respect to any Biogen Products developed and commercialized by Biogen pursuant to Section 8.3(e)(iv) following termination of this Agreement, as determined in accordance with Schedule C attached hereto.

"***Assigned Contract***" means the Contract relating to BIIB069 listed on Part 3 of Schedule A attached hereto to which Biogen or its Affiliates are bound including, without limitation, (a) all rights to receive payments under such Contract on and after the Effective Date, and (b) all of the claims or rights of action of Biogen or its Affiliates existing as of the Effective Date or arising after the Effective Date under such Contract.

"***Background Licensed Patent Rights***" means any Patent Rights that are (a) owned or Controlled by Biogen or its Affiliates as of the Effective Date and (b) actually used by Biogen or its Affiliates to manufacture BIIB069 prior to the Effective Date. For purposes of clarity, Background Licensed Patent Rights excludes the Acquired Patent Rights.

"***Background Sublicensed Intellectual Property"*** means the intellectual property rights relating to BIIB069 that are in-licensed by Biogen pursuant to the Retained Contracts.

"***BIIB069***" means the Antibody described on Schedule F attached hereto.

"***BLA"*** or "***Biologics License Application***" means a request for permission to introduce, or deliver for introduction, a biologic product into interstate commerce pursuant to the FDCA.

"***Business Day***" means a day other than Saturday, Sunday or any day on which commercial banks located in New York, New York are authorized or obligated by Law to close.

"***Calendar Quarter***" means the period beginning on the Effective Date and ending on the last day of the Calendar Quarter in which the Effective Date falls, and thereafter each successive period of three (3) consecutive calendar months ending on March 31, June 30, September 30 and December 31; provided, that, the final Calendar Quarter shall end on the last day of the Term.

"***Calendar Year***" means the period beginning on the Effective Date and ending on December 31 of the Calendar Year in which the Effective Date falls, and thereafter each successive period of twelve (12) months commencing on January 1 and ending on December 31; provided, that, the final Calendar Year shall end on the last day of the Term.

"***CDA***" means the Confidentiality Agreement dated as of November 19, 2015 (the "***CDA Effective Date***") by and between Kiniksa and Biogen.

"***Clinical Trial***" means, collectively, any Phase I Clinical Trial, Phase II Clinical Trial or Phase III Clinical Trial.

"***Combination Product***" means (a) any single product in finished form containing as active ingredients both (i) a Product and (ii) one or more other pharmaceutically-active compounds or substances; (b) any Product sold with another product(s) for a single invoice price; or (c) any Product sold as part of a bundle with other product(s) or service(s) (i.e., where a Product and such other product(s) or services are sold for a single invoice price or where a discount, rebate or other amount that reduces the price of a Product is provided in exchange for (or otherwise conditioned upon) the purchase of such other product(s) or services), to the extent not described in clause (a) or (b).

"***Commercialization***" or "***Commercialize***" means any and all activities directed to the offering for sale and sale of an Acquired Antibody or a Product including (a) activities directed to marketing, promoting, detailing, distributing, manufacturing, importing, selling and offering to sell that Acquired Antibody or Product; (b) interacting with Regulatory Authorities regarding the above; and (c) seeking pricing approvals and reimbursement approvals (as applicable) for that Product. When used as a verb, "***to Commercialize***" and "***Commercializing***" means to engage in Commercialization and "***Commercialized***" has a corresponding meaning.

"***Commercially Reasonable Efforts***" means with respect to the Development and Commercialization of an Acquired Antibody or Product by Kiniksa, the efforts and resources comparable to those undertaken by [***] in pursuing the development and commercialization of a compound that is of a similar market potential or profit potential and at a similar stage of development as such Acquired Antibody or Product. For purposes of the above, all relevant factors as measured by the facts and circumstances at the time such efforts are due shall be taken into account, including, as applicable and without limitation, mechanism of action; efficacy and safety; product profile; actual or anticipated Regulatory Authority approved labeling; the nature and extent of market exclusivity (including patent coverage, proprietary position and regulatory exclusivity), costs; time required for and likelihood of obtaining Marketing Authorization

(including reimbursement approval); the competitiveness of alternative products in the marketplace; and actual or projected profitability but excluding the effect of any consideration owed to Biogen or its Affiliates under this Agreement. Without limiting the foregoing, in circumstances where using Commercially Reasonable Efforts as defined above requires Kiniksa to take affirmative action, Kiniksa shall (a) assign responsibility for such obligations to specific employee(s) who are held accountable for progress and monitor such progress, each in a timely manner consistent with the nature of such obligations, (b) set and seek to achieve specific objectives for carrying out such obligations and (c) make and implement decisions and allocate resources designed to advance progress with respect to such objectives.

"***Completion of Technology Transfer***" has the meaning set forth in Schedule G attached hereto.

"***Contracts***" means any and all binding commitments, contracts, purchase orders, licenses, or other agreements, whether written or oral.

"***Control***" means, with respect to any Know-How or Patent Rights, the possession by a Party of the right to transfer or grant a license, sublicense or other rights to such Know-How or Patent Rights, as provided herein, without violating the terms of any agreement or arrangement with, infringing the Patent Rights of, or misappropriating the proprietary or trade secret information of, any Third Party and without violating any applicable Law.

"***Court***" means any court or arbitration tribunal of the United States, any domestic state, or any foreign country, and any political subdivision thereof.

"***Cover***" means, when referring to an Acquired Antibody or Product: (a) with respect to a patent, that, in the absence of a license granted to a Person under a claim included in such patent, the practice by such Person of a specified activity with respect to such Product or Acquired Antibody would infringe such claim (without regard to the validity or enforceability of such claim), or (b) with respect to a patent application, that, in the absence of a license granted to a Person under a claim included in such patent application, the practice by such Person of a specified activity with respect to such Product or Acquired Antibody would infringe such claim if such patent application were to issue as a patent.

"***Development***" or "***Develop***" means, with respect to any Acquired Antibody or Product, all non-clinical and clinical development activities with respect to such Acquired Antibody or Product that are undertaken by Kiniksa after the Effective Date, including formulation, process development, manufacturing scale-up, development-stage manufacturing, analytical method validation, manufacturing process validation, cleaning validation, quality assurance/quality control, statistical analysis, report writing, preclinical and clinical studies, clinical trial design and operations, clinical pharmacology studies, health economics and outcomes research studies, pharmacovigilance studies, the preparation and filing of Regulatory Filings and all regulatory affairs related to the foregoing. When used as a verb, "***Developing***" means to engage in

Development and "***Developed***" has a corresponding meaning.

"***Development Cost Breakeven Date***" means, with respect to any Biogen Product that is commercialized by Biogen after the effective date of termination pursuant to Section 8.3, the date on which Kiniksa has recouped from the payments made by Biogen pursuant to Section 8.3(e)(iv), an amount equal to (a) [***] plus (b) [***], plus (c) [***].

"***Development Costs***" means the aggregate out-of-pocket and internal costs incurred by Kiniksa, or for its account, determined in accordance with U.S. GAAP and the customary accounting principles of Kiniksa, consistently applied, that are allocable to the Development of an Acquired Antibody and/or Product.

"***Distributor***" means any Third Party which purchases its requirements for a Product in a country from Kiniksa or its Affiliates or licensees and is appointed as a distributor to distribute, market and resell such Product in such country, even if such Third Party is granted ancillary rights to develop, package or obtain regulatory approvals of such Product in order to distribute, market or sell such Product in such country.

"***Dollar***" means United States dollar, and "$" shall be interpreted accordingly.

"***EMA***" means the European Medicines Agency or any successor agency or authority thereto.

"***Encumbrance***" means any encumbrance, claim, mortgage, pledge, assessment, security interest, option, license, right of first refusal or preemptive right, hypothecation, equitable interest, preference, right of possession, deed of trust, lease, lien, levy, restriction on transferability, defect in title, charge or other encumbrance of any kind, whether voluntarily incurred or arising by operation of Law, any obligation to pay Taxes, any conditional sale or title retention agreement or other agreement granting any of the foregoing in the future or otherwise.

"***Exploit***" or "***Exploitation***" means to research, develop, make, have made, use, offer for sale, sell, import, export or otherwise exploit, or transfer possession of or title in, a compound or product.

"***FDA***" means the United States Food and Drug Administration or any successor agency or authority thereto.

"***FDCA***" means the United States Federal Food, Drug, and Cosmetic Act, as amended.

"***Field***" means any and all uses.

"***First Commercial Sale***" means, with respect to any Product in any country in the

Territory, the first sale, transfer or disposition for value to an end user of that Product in that country after Marketing Authorization for that Product has been received in that country; provided, that, the following shall not constitute a First Commercial Sale: (a) any sale to an Affiliate or licensee (unless the Affiliate or licensee is the last entity in the distribution chain of the Product), (b) any transfers of a Product without consideration or for nominal consideration for use in any Clinical Trial, or for any bona fide charitable, compassionate use or indigent patient program purpose where Products are sold at or below cost of goods sold or as a sample.

"***Force Majeure***" means any occurrence beyond the reasonable control of a Party that (a) prevents or substantially interferes with or delays the performance by such Party of any of its obligations hereunder and (b) occurs by reason of any act of God, flood, fire, explosion, earthquake, strike, lockout, labor dispute, casualty or accident, or war, revolution, civil commotion, act of terrorism, blockage or embargo, or any injunction, law, order, proclamation, regulation, ordinance, demand or requirement of any Governmental Entity or of any subdivision, authority or representative of any such Governmental Entity.

"***FTE***" means the equivalent of the work of one employee full time for one year consisting of at least a total of 45.5 weeks or 1,820 hours per year (excluding vacations and holidays). For purposes of clarity, no one person shall be permitted to account for more than one FTE.

"***FTE Rate***" means [***] per FTE per year.

"***GLP*** *or* "***Good Laboratory Practice***" means all applicable then-current standards for laboratory activities for pharmaceuticals, as set forth in the FDA's Good Laboratory Practice regulations as defined in 21 C.F.R. Part 58 or the Good Laboratory Practice principles of the Organization for Economic Co-Operation and Development (OECD), and such standards of good laboratory practice as are required by the European Union and other organizations and governmental agencies in countries in which a Product is intended to be sold, to the extent such standards are not less stringent than United States Good Laboratory Practice.

"***GMP***" or "***Good Manufacturing Practice***" means all applicable then-current standards for manufacturing, including, as applicable, (a) the principles detailed in the U.S. Current Good Manufacturing Practices, 21 C.F.R. §§ 201, 211, 600 and 610 and all applicable FDA guidelines and requirements, (b) European Directive 2003/94/EC for medicines and investigational medicines for human use and the applicable guidelines stated in the Eudralex guidelines, (c) the principles detailed in the applicable ICH guidelines, (d) the conduct of an inspection by a Qualified Person and the execution by such Qualified Person of an appropriate certification of inspection; and (e) the equivalent applicable Law in any relevant country, each as may be amended and applicable from time to time.

"***Governmental Entity***" means any court, tribunal, arbitrator, Regulatory Authority, agency, commission, department, ministry, official or other instrumentality of the United States or other country, or any supra-national organization, or any foreign or domestic, state, county,

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city or other political subdivision.

"***IND***" means (a) an Investigational New Drug Application (as defined in the FDCA and the regulations promulgated thereunder) or any successor application or procedure required to initiate clinical testing of a therapeutic product in humans in the United States; (b) the equivalent of an Investigational New Drug Application that is required in any other country or region before beginning clinical testing of a therapeutic product in humans in such country or region (including any Clinical Trial Authorization ("***CTA***") required to initiate clinical testing of a therapeutic product in humans in the United Kingdom); and (c) all supplements and amendments to any of the foregoing.

"***Indemnifying Party***" means the Biogen Indemnifying Party or the Kiniksa Indemnifying Party, as the case may be.

"***Indemnified Party***" means the Biogen Indemnified Party or the Kiniksa Indemnified Party, as the case may be.

"***Indication***" means any human indication, disease or condition in the Field, which can be treated, prevented, cured or the progression of which can be delayed, excluding an expansion of label claim for an already approved indication. For example, [***]: (a) [***], or (b) [***].

"***Initiation***" means with respect to a Clinical Trial, the first date that a subject (healthy volunteer or patient) is first dosed in such Clinical Trial.

"***Inventory***" means the inventory of BIIB069 listed as Inventory in Part 4 of Schedule A attached hereto.

"***Kiniksa Know-How***" means any Know-How, other than the Acquired Know-How and the Know-How included in the Background Sublicensed Intellectual Property, that is owned or Controlled by Kiniksa and which relates to, or is used by Kiniksa in connection with, the Development and Commercialization, including the manufacture, use, offer for sale, sale or importation, of any Acquired Antibody or Product.

"***Kiniksa Patent Rights***" means any Patent Rights, other than the Acquired Patent Rights, the Background Licensed Patent Rights and the Patent Rights included in the Background Sublicensed Intellectual Property, that are owned or Controlled by Kiniksa that contain one or more claims that Cover any Acquired Antibody or Product, including the Development, Commercialization, manufacture, use, offer for sale, sale or importation of any such Acquired Antibody or Product.

"***Kiniksa Third Party Agreement***" means any agreement by and between Kiniksa and any Third Party pursuant to which Kiniksa Controls any Kiniksa Know-How and/or Kiniksa Patent Rights.

"***Know-How***" means, collectively, any knowledge, information, techniques, technology, trade secrets, inventions (whether patentable or not), discoveries, methods, know-how, data and results (including complementarity determining region (CDR) sequence information and pharmacological and toxicological data and results), analytical and quality control data and results, regulatory documents, and other information, compositions of matter, cells, cell lines, assays, animal models and other physical, biological, or chemical material.

"***Knowledge***" means, with respect to Biogen, the actual knowledge of the individuals listed on Schedule H attached hereto.

"***Law***" means any federal, state, local or foreign law, statute, code or ordinance, or any rule or regulation promulgated by any Governmental Entity including all decisions of any Courts having the effect of law in each such jurisdiction.

"***Liability***" means any and all debts, liabilities and obligations, whether known or unknown, asserted or unasserted, determinable or otherwise, accrued or fixed, absolute orcontingent, liquidated or unliquidated, incurred or consequential, or matured or unmatured, including, without limitation, those arising under any Law, Litigation, Order, or Contract.

"***Litigation***" means any suit, action, arbitration, cause of action, claim, complaint, criminal prosecution, investigation, inquiry, demand letter, judicial, arbitration or other administrative proceeding, whether at law or at equity, before or by any Court, Governmental Entity, arbitrator or other tribunal.

"***Marketing Authorization***" means, with respect to a Product, the regulatory approval required by applicable Law to sell such Product in a country or region in the Territory. For purposes of clarity, (a) "***Marketing Authorization***" in the United States means final approval of an NDA, sNDA or BLA permitting marketing of such Product in interstate commerce in the United States; and (b) "***Marketing Authorization***" in Europe means marketing authorization for such Product granted either by a Regulatory Authority in any country in Europe or by the EMA pursuant to Council Directive 2001/83/EC, as amended, or Council Regulation 2309/93/EEC, as amended.

"***NDA***" means a New Drug Application, as defined in the FDCA and regulations promulgated thereunder or any successor application or procedure required to sell any Product in the United States.

"***Net Sales***" means, with respect to a Product in any country in the Territory, the gross amount invoiced by Kiniksa, its Affiliates, or licensees for the sale or other disposition of such Product in such country to Third Parties, including Distributors ("**Gross Sales**"), less the following deductions (such deductions, collectively, "**Sales Returns and Allowances**"):

(a) sales returns and allowances actually paid, granted or accrued on the Product, including trade, quantity, prompt pay and cash discounts and any other adjustments, including

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those granted on account of price adjustments or billing errors;

(b) credits or allowances given or made for rejection, recall, return or wastage replacement of Product or for rebates or retroactive price reductions (including Medicare, Medicaid, managed care and similar types of rebates and chargebacks);

(c) taxes, duties or other governmental charges levied on or measured by the billing amount for such Product, as adjusted for rebates and refunds, but which shall not include any tax, duty, or other charge imposed on or measured by net income (however denominated) or any franchise taxes, branch profits taxes, or similar tax; and

(d) charges for freight, customs and insurance directly related to the distribution of the Product and wholesaler and Distributor administration fees;

in each case, to the extent such deductions: (i) are reasonable and customary, (ii) included in the gross invoiced sales price for the Product or otherwise directly paid, allowed, accrued, or incurred by such Party, its Affiliates or licensees with respect to the sale of such Product (iii) applicable and in accordance with standard allocation procedures, (iv) have not already been deducted or excluded, (v) are incurred in the ordinary course of business in type and amount consistent with good industry practice, and (vi) are determined in accordance with, and as recorded in revenues under, US GAAP.

For purposes of clarity, (1) Net Sales shall not be imputed to transfers of Product (i) without consideration or for nominal consideration for use in any Clinical Trial or any other human studies reasonably necessary to comply with any applicable Law or regulation or any request by a Regulatory Authority, (ii) for any bona fide charitable, compassionate use or indigent patient or other similar program purpose where Products are sold at or below cost of goods sold, or (iii) in commercially reasonable quantities as samples for promotional purposes; (2) in the case of any transfer of any Product between or among Kiniksa and its Affiliates or licensees for resale, Net Sales shall be determined based on the sale made by such Affiliate or licensee to a Third Party (including any Distributors).

Notwithstanding the foregoing, in the event a Product is sold as a component of a Combination Product in any country in the Territory in any Calendar Quarter, Net Sales shall be calculated by multiplying the Net Sales of the Combination Product (calculated in the same manner as set forth above as if the Combination Product were a Product) in such country during such Calendar Quarter by the fraction A/(A+B), where A is the average Net Sales of the Product when sold separately in such country during such Calendar Quarter and B is the average Net Sales of the other pharmaceutically active compounds or substances included in the Combination Product (calculated in the same manner as set forth above as if the other pharmaceutically active compounds or substances were a Product) when sold separately in such country during such Calendar Quarter. In the event that no separate sales of the Product or any other pharmaceutically active compounds or substances included in a Combination Product are made by Kiniksa, its Affiliates or licensees in a country during a Calendar Quarter in which such

Combination Product is sold in such country, the average Net Sales in the above described equation shall be replaced with reasonable good faith estimate of the fair market value, as mutually determined by the Parties, of the Product and each of the other pharmaceutically active compounds or substances included in such Combination Product.

"***Order***" means any judgment, order, writ, injunction, ruling, stipulation, determination, award or decree of or by, or any settlement under the jurisdiction of, any Court or Governmental Entity.

"***OSMR***" means the oncostatin M receptor, one of the receptor proteins for oncostatin M that in humans is encoded by the OSMR gene.

"***Patent Rights***" means the rights and interests in and to issued patents and pending patent applications (which, for purposes of this Agreement, include certificates of invention, applications for certificates of invention and priority rights) in any country or region, including any divisionals, continuations, continuations-in-part, substitutions, patents of addition, reissues, extensions, re-examinations or renewal applications related to, or claiming priority to, the foregoing (including any supplemental patent certificates) or any confirmation patent or registration patent, and all patents issuing on, and all foreign counterparts of, any of the foregoing.

"***Permitted Encumbrances***" means (a) statutory liens with respect to the payment of Taxes, in all cases which are not yet due or payable; and (b) statutory liens of landlords, suppliers, mechanics, carriers, materialmen, warehousemen, service providers or workmen and other similar liens imposed by applicable Law created in the ordinary course of business and which liens (i) do not constitute a default or breach under the Assigned Contract and (ii) have not had, and would not reasonably be expected to have, individually or in the aggregate, a material adverse effect on the Purchased Assets.

"***Person***" means any natural person, corporation, general partnership, limited partnership, limited liability company, proprietorship, joint venture, other business organization, trust, entity, union, association or Governmental Entity.

"***Phase I Clinical Trial***" means a human clinical trial for any Product in any country that would satisfy the requirements of 21 CFR 312.21(a). For clarity, a Phase Ia or Phase Ib clinical trial shall be classified as a "Phase I Clinical Trial."

"***Phase II Clinical Trial***" means a human clinical trial conducted in any country that would satisfy the requirements of 21 CFR 312.21(b) and is intended to explore one or more doses, dose response, and duration of effect, and to generate initial evidence of clinical activity and safety, for any Product in the target patient population. For clarity, a Phase IIa or Phase IIb clinical trial shall be classified as a "Phase II Clinical Trial."

"***Phase III Clinical Trial***" means a clinical trial in an extended human patient population

designed to obtain data determining efficacy and safety of any Product to support Marketing Authorizations in the proposed therapeutic Indication, as more fully defined in 21 C.F.R. §312.21(c), or its successor regulation, or the equivalent in any foreign country. For clarity, a Phase IIIa or Phase IIIb clinical trial shall be classified as a "Phase III Clinical Trial."

"***Product***" means any product (a) that contains or incorporates any Acquired Antibody or (b) the manufacture, use or sale of which is Covered by a Valid Claim.

"***Product Trademarks***" means any trademark used by Kiniksa in connection with the Commercialization of any Product.

"***Regulatory Authority***" means any national, supra-national, regional, state or local regulatory agency, department, bureau, commission, council or other governmental entity with authority over the distribution, importation, exportation, manufacture, production, use, storage, transport, clinical testing, marketing, pricing or sale of a Product, including the FDA, the EMA, and the European Commission.

"***Regulatory Filing***" means, collectively: (a) any IND, CTA, MAA, BLA, establishment license application, drug master file, application for designation as an "Orphan Drug" under the Orphan Drug Act, for "Fast Track" status under Section 506 of the FDCA (21 U.S.C. § 356) or for a Special Protocol Assessment under Section 505(b)(4)(B) and (C) of the FDCA (21 U.S.C. § 355(b)(4)(B)) and all other similar filings (including counterparts of any of the foregoing in any country or region in the Territory); (b) all supplements and amendments to any of the foregoing; and (c) all data and other information contained in, and correspondence relating to, any of the foregoing.

"***Retained Contracts***" means the Contracts listed on Schedule E attached hereto.

"***Royalty Term***" means with respect to each Product, the period beginning on the date of First Commercial Sale of such Product in any country in the Territory and ending on the latest of (a) the expiration of the last to expire Valid Claim that Covers the composition of matter, manufacture, use or sale of such Product in such country, (b) the expiration of regulatory exclusivity in such country, and (c) ten (10) years from the date of the First Commercial Sale of such Product in such country.

"***Tax***" *or* "***Taxes***" means all income, excise, gross receipts, ad valorem, sales, use, employment, environmental, franchise, profits, gains, property, transfer, value added, payroll, escheat or abandoned property, intangibles or other taxes, fees, stamp taxes, duties, charges, levies or assessments of any kind whatsoever (whether payable directly or by withholding), together with any interest and any penalties, additions to tax or additional amounts imposed by any Governmental Entity with respect thereto, whether as a primary obligor, as a result of being a transferee, successor or a member of an affiliated, consolidated, unitary, combined or other group, by contract, pursuant to Law or otherwise.

"***Territory***" means worldwide.

"***Third Party***" means a Person other than Kiniksa, Biogen or their respective Affiliates.

"***Third Party Intellectual Property***" means any intellectual property rights generated prior to the Effective Date under a Contract between Biogen or its Affiliates and a Third Party in the Exploitation of the Acquired Antibody that are not Controlled by Biogen or its Affiliates.

"***US GAAP***" means United States Generally Accepted Accounting Principles.

"***Valid Claim***" means (a) an issued and unexpired patent claim within the Acquired Patent Rights, the Kiniksa Patent Rights, the Background Licensed Patent Rights or the Background Sublicensed Intellectual Property; or (b) a claim of a pending patent application within the Acquired Patent Rights, the Kiniksa Patent Rights, the Background Licensed Patent Rights or the Background Sublicensed Intellectual Property and that, in the case of any such patent application, was filed in good faith, has not been pending for more than [***]) years, and has not been abandoned or finally disallowed.

Additional Definitions. In addition, each of the following definitions shall have the respective meanings set forth in the section of this Agreement indicated below:

Definition	Section
Agreement	Preamble
Amgen	2.5.2
Amgen Agreement	2.5.2
Annual [***]	3.3.2
Assigned Kiniksa Agreements	8.3(e)(ii)
Assumed Liabilities	2.3
ATCC Agreement	Schedule E
Bioequivalence	5.3.2(c)
Bioequivalent	5.3.2(c)
Biogen	Preamble

Biogen Indemnified Parties	9.2
Biogen Product	8.3(e)(iv)
CDA Effective Date	CDA definition
Change of Control Transaction	3.4.2
Claims	9.2
Competing Drug	5.3.2(c)
Confidential Information	7.1
CTA	IND definition
Data Package	3.4.1(b)
Diligence Period	3.4.1(c)
Effective Date	Preamble

Exchange Act	3.4.2
Excluded Assets	2.2
Excluded Liabilities	2.4
Exclusivity Period	2.7
Exclusivity Term	3.4.1(d)
Exempt Transaction	3.4.3
Gross Sales	Definition of Net Sales
ICH	3.2
Infringement	4.2.1
Infringement Notice	4.2.1
Infringement Response	4.2.2
Issuing Party	7.5.2
Kiniksa	Preamble
Kiniksa Election Notice	8.4
Kiniksa Indemnified Parties	9.3
Kiniksa Supply Agreement	8.3(e)(vi)
Losses	9.2
Negotiation Notice	3.4.1(b)
Non-Assignable Kiniksa Agreements	8.3(e)(ii)
Notice Review Period	3.4.1(b)
Party/Parties	Preamble
Patent Assignment Agreement	2.6.2
Purchased Assets	2.1
Recovery	4.2.5
Release	7.5.2
Report	3.3.1
Retained Contract Payments	5.4
Reviewing Party	7.5.2
Right of First Negotiation	3.4
ROFN Transaction	3.4.1(a)
Sales Returns and Allowances	Definition of Net Sales
Sigma Agreement	Schedule E
Technology Transfer Fees	5.1.2
Term	8.1
Transfer Taxes	5.8.2
Trigger Notice	3.4.1(a)
Unsolicited Offer	3.4.1(b)
Upfront Fee	5.1.1

1.2 **Construction of Certain Terms and Phrases**. Unless the context of this Agreement otherwise requires: (a) words of any gender include each other gender; (b) words using the singular or plural number also include the plural or singular number, respectively; (c) the terms "hereof," "herein," "hereby" and derivative or similar words refer to this entire

Agreement; (d) the terms "Article" or "Section" refer to the specified Article or Section of this Agreement; (e) the term "or" has, except where otherwise indicated, the inclusive meaning represented by the phrase "and/or"; and (f) the term "including" means "including without limitation." Whenever this Agreement refers to a number of days, such number shall refer to calendar days unless Business Days are specified.

2. PURCHASE AND SALE OF ASSETS

2.1 **Purchase and Sale of Assets**. Upon the terms and subject to the conditions set forth in this Agreement, Biogen hereby sells, conveys, assigns, transfers and delivers to, and shall cause its Affiliates to sell, convey, assign, transfer and deliver to, Kiniksa, and Kiniksa hereby purchases and acquires from each of Biogen or its Affiliates, as the case may be, all of Biogen's and its Affiliates' right, title and interest in and to the following assets described or set forth on Schedule A attached hereto (collectively, the "***Purchased Assets***") free and clear of all Encumbrances (other than Permitted Encumbrances):

(a) the Acquired Patent Rights;

(b) the Acquired Know-How;

(c) the Assigned Contracts; and

(d) the Inventory.

2.2 **Excluded Assets**. Notwithstanding the provisions of Section 2.1, no right, title or interest is being sold, assigned, transferred, conveyed or delivered to Kiniksa in or to (a) any of the property and assets of Biogen that are not listed on Schedule A or (b) any rights or claims of Biogen under this Agreement (collectively, the "***Excluded Assets***").

2.3 **Assumed Liabilities**. Subject to the terms and conditions of this Agreement, on and after the Effective Date, Kiniksa shall assume and agree to pay, perform and discharge the following Liabilities of Biogen (the "***Assumed Liabilities***"):

(a) all Liabilities and obligations resulting from the ownership, use, operation or maintenance of the Purchased Assets and/or the Exploitation of any Acquired Antibody and/or Product, by Kiniksa to the extent that such Liability arises from any event, condition or circumstance occurring after the Effective Date and not resulting from any breach by Biogen of any of its obligations under this Agreement;

(b) all Liabilities arising under the Assigned Contracts after the Effective Date to the extent that such Liabilities are not attributable to any failure by Biogen or any of its Affiliates to comply with the terms thereof prior to the Effective Date;

(c) all Liabilities for Transfer Taxes described in Section 5.8.2; and

all Taxes imposed on the Purchased Assets or that otherwise arise with respect to the use of the Purchased Assets, in each case, for any taxable period (or portion thereof) beginning after the Effective Date.

2.4 **Excluded Liabilities**. Biogen shall retain, and shall be responsible for paying, performing and discharging when due, and Kiniksa shall not assume or have any responsibility for paying, performing or discharging, any Liabilities of Biogen and its Affiliates other than the Assumed Liabilities (the "***Excluded Liabilities***"). Without limiting the foregoing, neither Kiniksa nor its Affiliates shall be obligated to assume, and neither of them does assume, and each of them hereby disclaims responsibility for, any of the following Liabilities of Biogen and its Affiliates:

(a) any Liability attributable to any asset, property or right that is not included in the Purchased Assets;

(b) any Liability attributable to the research, development or other activity conducted by Biogen or any Affiliate related to the Acquired Antibody on or prior to the Effective Date;

(c) all Liabilities arising under the Assigned Contracts prior to the Effective Date to the extent that such Liabilities are not attributable to any failure by Kiniksa or any of its Affiliates to comply with the terms thereof after the Effective Date; and

(d) all Taxes imposed on the Purchased Assets or that otherwise arise with respect to the use of the Purchased Assets, in each case, for any taxable period (or portion thereof) ending on or prior to the Effective Date; all Taxes of Biogen or any of its Affiliates that are or may become payable with respect to all taxable periods, including any Liability for such Taxes that arise as a result of the transactions contemplated by this Agreement but excluding any Transfer Taxes described in Section 5.8.2; and, except as otherwise provided in Section 5.8.3, all Taxes required to be withheld or deducted by applicable Law in connection with the transactions contemplated by this Agreement.

2.5 License Grants; Obligations of Kiniksa Under Retained Contracts.

2.5.1 Grant of License/Sublicense by Biogen to Kiniksa. Biogen hereby grants, on behalf of itself and its Affiliates, to Kiniksa a non-exclusive, sublicensable (through multiple tiers of sublicensees, but subject to Section 3.4), license under the Background Licensed Patent Rights and sublicense under the Background Sublicensed Intellectual Property, in each case, for the Exploitation of Acquired Antibodies and/or Products for use in the Field and in the Territory.

2.5.2 Grant of License by Kiniksa to Biogen. Kiniksa hereby grants, on behalf of itself and its Affiliates, to Biogen a worldwide, non-exclusive, fully paid, royalty-free, sublicensable (through multiple tiers of sublicensees), perpetual license (which is irrevocable,

and not subject to termination for any reason) under the Acquired Patent Rights in the Territory, wherein such license is limited to making, using or selling any Antibody or antigen binding portion of an Antibody that [***] OSMR. Kiniksa acknowledges that Biogen and/or its Affiliates is/are obligated under the Asset Purchase Agreement between Amgen Inc. ("***Amgen***") and Biogen, dated as of August 12, 2013 (the "***Amgen Agreement***"), to sublicense such Acquired Patent Rights to Amgen.

2.5.3 Obligations of Kiniksa Under Retained Contracts. Kiniksa hereby agrees to be bound by and comply with, and agrees to cause its Affiliates and sublicensees to be bound by and comply with, all of the terms, conditions, obligations, and any restriction of rights, applicable to a sublicensee of Biogen under either of the Retained Contracts. Without limiting the foregoing, Kiniksa hereby agrees as follows in connection with the exercise of its rights and the performance of its obligations under this Agreement:

(a) Specific Obligations of Kiniksa under ATCC Agreement.

(i) All capitalized terms used in this Section 2.5.3(a) and not otherwise defined shall have the respective meanings set forth in the ATCC Agreement.

(ii) In accordance with Section 8.3 of the ATCC Agreement, Kiniksa shall comply with, and shall contractually obligate its Related Parties to comply with, all United States laws and regulations controlling the export and re-export of certain commodities and technical data, including without limitation, all Export Administration Regulations of the United States Department of Commerce (as presently promulgated or hereinafter modified or amended).

(iii) In accordance with Section 8.4 of the ATCC Agreement, Kiniksa shall obtain, and shall contractually obligate its Related Parties to obtain, all authorities, consents and clearances required for the purchase, importation, exportation transportation, distribution, demonstration and Sale of Products under this Agreement that are Biogen Products for purposes of the ATCC Agreement to and within the Territory. Kiniksa shall further comply with, and shall contractually obligate its Related Parties to comply with, all applicable foreign and domestic, federal, state and local statutes, ordinances and regulations in connection with its purchase, importation, exportation transportation, distribution, demonstration and Sale of Products under this Agreement that are Biogen Products for purposes of the ATCC Agreement.

(iv) To the extent Kiniksa uses any Biological Materials in the Development and/or Commercialization of any Acquired Antibody or Product under this Agreement, Kiniksa certifies, and shall contractually obligate its Related Parties to certify, in accordance with Section 11.1 of the ATCC Agreement, that Kiniksa and its Related Parties, as applicable, shall:

(A) ensure that only qualified personnel work with such Biological Material in proper facilities;

(B) provide sufficient internal security to assure access to such Biological Material only by those individuals authorized to work with them;

(C) not transfer, export, resell, or otherwise dispose of any such Biological Material to any Third Parties under any circumstances without written authorization by Biogen and ATCC and the appropriate government agencies or as explicitly provided for in the ATCC Agreement;

(D) not permit access to such Biological Materials by foreign entities or individuals when to do so would be in violation of export control laws;

(E) comply with all applicable federal, state or local laws and regulations pertaining to such Biological Material or their handling, storage, use transportation; and

(F) unless requested otherwise by Biogen or ATCC, destroy all such Biological Material according to accepted practices for destruction of biohazardous material upon completion of work or expiration or termination of this Agreement or a subsequent license with Biogen for the ATCC Materials, whichever occurs first, as set forth in ARTICLE 12 of the ATCC Agreement.

(b) Specific Obligations of Kiniksa under Sigma Agreement.

(i) All capitalized terms used in this Section 2.5.3(b) and not otherwise defined shall have the respective meanings as set forth in the Sigma Agreement.

(ii) Kiniksa hereby agrees that it shall be bound by the terms of the Sigma Agreement set forth on Schedule B attached hereto.

(iii) Kiniksa hereby further agrees, in accordance with Section 2.2 of the Sigma Agreement, that each sublicense granted by Kiniksa or its Affiliates, their sublicensees or their further sublicensees (whether direct or indirect) shall include the first two sentences of Section 2.5 of the Sigma Agreement and the terms and conditions set forth in Exhibit B of the Sigma Agreement (each of which is set forth on Schedule B attached hereto).

2.6 Support by Biogen.

2.6.1 Technology Transfer. Biogen shall transfer to Kiniksa the Acquired Know-How and the Inventory listed on Parts 2 and 4 of Schedule A, respectively, in accordance with the protocols and timeframes listed on Schedule G attached hereto. The Completion of Technology Transfer for purposes of Schedule G shall be deemed to have occurred on the first anniversary of the Effective Date, unless the Parties otherwise agree that such transfer is completed upon an earlier date (as determined by the completion of activities provided on such Schedule G). Subject to Section 2.6.4, to the extent reasonably requested by Kiniksa, [***] at any time prior to the [***] anniversary of the Effective Date, Biogen shall provide reasonable consulting support to Kiniksa in connection with its Exploitation of Products. Kiniksa acknowledges that (a) any materials comprising Inventory transferred by Biogen to Kiniksa under this Agreement are experimental in nature and may have unknown characteristics and therefore agrees to use prudence and reasonable care in the use, handling, storage, transportation and disposition and containment of any such materials and (b) if Kiniksa chooses to use such materials in any human application, including in the conduct of any Clinical Trial, it shall do so at its own risk.

2.6.2 Patent Rights Transfer. Promptly (and in no event later than thirty (30) days) following the Effective Date, Biogen shall provide Kiniksa, or Kiniksa's designated attorneys, with copies of the file histories and supporting data of the pending patent application (provisional and otherwise) within the Acquired Patent Rights in Biogen's possession, and shall promptly take any actions necessary to obtain and provide to Kiniksa, or Kiniksa's designated attorneys, copies of any such file histories not in Biogen's possession; provided, that, Biogen shall use reasonable efforts to ensure that such copies of the file histories will be complete. Additionally, Biogen shall, from time to

time, take such actions as are reasonably requested by Kiniksa to perfect the transfer of Biogen's right, title and interest in the Acquired Patent Rights to Kiniksa, including the execution of (a) any documents needing inventor signature, (b) the patent assignment agreement attached hereto as Exhibit A (the "***Patent Assignment Agreement***") and (c) any other patent assignments that may be reasonably required in any other jurisdiction.

2.6.3 Other Cooperation. Subject to Section 2.6.4, Biogen agrees to use reasonable efforts to (a) make its employees, agents and consultants reasonably available to, and at the expense of, Kiniksa (or to Kiniksa's authorized attorneys, agents or representatives), and provide contact information in Biogen's possession and control with respect to the listed inventors of the Acquired Patent Rights, to the extent, in any case, reasonably necessary to enable Kiniksa or its designees to undertake preparation of U.S. and foreign applications claiming priority to Acquired Patent Rights and prosecution and maintenance of such applications.

2.6.4 Cost of Biogen Support. The Parties hereby agree that the consulting support and other cooperation activities provided by Biogen under Sections 2.6.1 and 2.6.3 shall be provided at Biogen's sole expense for up to [***] hours of such consulting support and cooperation activities and thereafter at Kiniksa's sole expense (including Biogen's employee

costs at the FTE Rate).

2.6.5 Maintenance of Retained Contracts. Biogen agrees that it will not, and will ensure that its Affiliates do not, without Kiniksa's prior written consent (a) sell, assign, transfer, convey, deliver or otherwise divest its interests in any of the Retained Contracts to a Third Party, (b) mortgage or otherwise encumber its interests in any of the Retained Contracts, in a manner that adversely affects, or would reasonably be expected to adversely affect, Kiniksa's rights or obligations under this Agreement, (c) amend any of the Retained Contracts in a manner that adversely affects the rights granted to Kiniksa under this Agreement or (d) undertake any action that would constitute a material breach of, and allow the Third Party that is a party to any Retained Contract to terminate, any Retained Contract.

2.6.6 Completeness of Patent Rights and Know-How. Except as set forth in Schedule I attached hereto, Biogen agrees that, if at any time after the Effective Date, Biogen becomes aware (including as a result of written notice from Kiniksa) and determines that any Patent Rights or Know-How that (a) were owned or Controlled by Biogen as of the Effective Date and used by Biogen in, and necessary for, the Exploitation of BIIB069 as it existed as of the Effective Date were not included in the Acquired Patent Rights or Acquired Know-How or (b) were owned by Biogen or licensed to Biogen as of the Effective Date as part of the Background Licensed Patent Rights or Background Sublicensed Intellectual Property, as applicable, and were not included in the license grant to Kiniksa in Section 2.5.1, then Biogen shall promptly notify Kiniksa of such determination. Biogen shall promptly take such actions as may be reasonably necessary to deliver such Patent Rights or Know-How, as applicable, to Kiniksa, in a manner consistent with the assignment and delivery terms of this Agreement applicable to Acquired Patent Rights or Acquired Know-How, as the case may be, or license such Patent Rights or Know-How to Kiniksa as Background Licensed Patent Rights or Background Sublicensed Intellectual Property, as the case may be, in a manner consistent with Section 2.5.1.

2.7 **Exclusivity**. In consideration of the transactions contemplated hereby, during the period beginning on the Effective Date and, subject to Section 8.3(b) of this Agreement, continuing until [***] (the "***Exclusivity Period***"), neither Kiniksa nor any of its Affiliates shall, directly or indirectly, (i) conduct any activity, either on its own or for its benefit, or with, for the benefit of, or sponsored by, any Third Party, or grant any license to any Third Party to utilize any Know-How or Patent Rights owned or controlled by Kiniksa or any of its Affiliates, that, in any case, involves the identification, generation, research, development, manufacture, commercialization, sales, marketing, promotion or distribution of any compound or biologic that [***] or (ii) appoint, grant any right or license to or otherwise authorize any Third Party to perform any of the foregoing activities, other than, in any such case, the Development and Commercialization of Acquired Antibodies and Products pursuant to this Agreement (whether by sale, license, sublicense or other transfer).

3. DEVELOPMENT AND COMMERCIALIZATION OF PRODUCTS

3.1 **Responsibility**. Kiniksa shall have the sole right and responsibility, at its sole cost and expense, for the conduct of all Development and Commercialization activities applicable to Acquired Antibodies and Products for use in the Field and in the Territory after the Effective Date, including without limitation, (a) all pre-marketing, marketing, promotion, sales, distribution, import and export activities (including securing reimbursement, sales and marketing and conducting any post-marketing trials or databases and post-marketing safety surveillance); (b) making all Regulatory Filings for any Acquired Antibody and/or Product and otherwise seeking all Marketing Authorization for any Product within the Territory, as well as all correspondence and communications with Regulatory Authorities regarding such matters; (c) conducting all manufacturing development and/or manufacturing activities with respect to the Acquired Antibodies and Products; (d) reporting of all adverse events to Regulatory Authorities if and to the extent required by applicable Law; (e) submitting applications for reimbursement with respect to any Product in any country in the Territory and (f) booking all sales of Products in the Territory.

3.2 **Diligence**. Kiniksa shall use Commercially Reasonable Efforts to Develop and Commercialize all Acquired Antibodies and Products and to commit such resources (including employees, consultants, contractors, facilities, equipment and materials) as are necessary to conduct such Development and Commercialization activities. Kiniksa shall perform its obligations under this Agreement in good scientific manner and in compliance with all applicable Law. For purposes of clarity, with respect to each Development and/or Commercialization activity that will or would reasonably be expected to be submitted to a Regulatory Authority in support of a Regulatory Filing or Marketing Authorization, Kiniksa shall comply in all material respects with GLPs, GMPs or Good Clinical Practices (or, if and as appropriate under the circumstances, International Council for Harmonisation ("**ICH**") guidance or other comparable regulation and guidance of any Regulatory Authority in any country or region in the Territory).

3.3 Reports.

(b) Records; Reports. Kiniksa shall (a) maintain records of its Development and Commercialization activities with respect to Acquired Antibodies and Products under this Agreement in sufficient detail and in good scientific manner, which shall reflect work performed and results achieved in the conduct of such Development and Commercialization activities and keep Biogen reasonably informed regarding the Development and Commercialization activities conducted with respect to Acquired Antibodies and Products by providing Biogen with reports at least [***] summarizing the activities undertaken by Kiniksa, its Affiliates and its licensees for the relevant [***] period (each, a "***Report***").

3.3.1 Content of Reports. Any Reports provided pursuant to Section 3.3.1 will include at least information regarding: (a) completed activities with respect to the Development of Acquired Antibodies and Products as well as the anticipated Development activities planned in the subsequent [***]; (b) activities with respect to the milestone events described in Section 5.2 including, when such milestone events are expected to be achieved and whether or not such

milestone events have been achieved; (c) an updated list of the Acquired Patent Rights or Kiniksa Patent Rights; and (d) the anticipated date and actual date, as applicable, of the First Commercial Sale of each Product in each country of the Territory; provided, however, that after a Product receives Marketing Authorization, the information required in (a) will only need to be provided [***] and will include planned activities for the subsequent [***]. In addition, in order to enable Biogen to prepare its quarterly and annual public disclosures regarding Biogen's results of operations, on a Product- by-Product basis, upon the earlier of (i) [***] year prior to the anticipated First Commercial Sale of such Product in any country in the Territory, or (ii) the date of Kiniksa's submission of a Regulatory Filing for a Product in such country, and on a [***] basis thereafter, Kiniksa shall prepare a commercialization report, which report shall include a timeline for achieving First Commercial Sale, a non-binding [***] good faith rolling forecast of Gross Sales and Net Sales of Products in the Field in the Territory, broken down by Calendar Quarters (such forecast, an "***Annual [***]***"). Thereafter, Kiniksa shall provide to Biogen an updated Annual [***] each Calendar Year of the Term.

3.4 **Right of First Negotiation**. Biogen shall have a right of first negotiation, as provided in this Section 3.4, to negotiate with Kiniksa for an agreement providing for the grant to Biogen or its Affiliates of the right to Exploit any Product in the Field and in the Territory under the circumstances described below (each, a "***Right of First Negotiation***").

3.4.1 Procedure.

(a) If Kiniksa and/or an Affiliate determines to seek or seeks to (including without limitation by determination of its Board of Directors or management and/or through the commencement of negotiations), directly or indirectly through Kiniksa and/or an Affiliate, (i) grant a license or sublicense to a Third Party to Exploit any Product, or (ii) assign, transfer or sell to any Third Party all or any portion of its rights to Exploit any Product, including through a Change of Control Transaction (as defined in Section 3.4.2 below), but excluding any Exempt Transaction (as defined in Section 3.4.3 below) (each transaction described in (i) or (ii) above, a "***ROFN Transaction"***), then Kiniksa will notify Biogen in writing which notice shall include a description of the assets or products that are the subject of the ROFN Transaction and provide to Biogen (A) a confidential summary of the Product and any other products and programs that are part of the ROFN Transaction; provided, that, solely to the extent that (y) the ROFN Transaction is with respect to a Change of Control Transaction involving an Unsolicited Offer (as defined below) and (z) [***] of any other products and programs of Kiniksa (other than the Product) if the Board of Directors or similar governing body of Kiniksa determines in good faith, after consultation with outside counsel, that such action would be [***], (B) the intended scope, if applicable (i.e., field and territory), and form of the ROFN Transaction and (C) the Notice Review Period applicable to such ROFN Transaction as provided in Section 3.4.1(b)(i) or (ii) below (each a "***Trigger Notice***").

(b) If Biogen desires to evaluate a ROFN Transaction after receipt of the Trigger Notice, Biogen will provide Kiniksa with a written notice (each, a "***Negotiation Notice***") as soon as reasonably possible but not longer than (i) within [***] days after Biogen's

receipt of the Trigger Notice if the Trigger Notice is with respect to a Change of Control Transaction with a Third Party that provided Kiniksa with a written *bonafide*, arms- length, unsolicited offer (which was not directly or indirectly solicited or induced by Kiniksa or its employees, directors, agents or representatives) (each an "***Unsolicited Offer***"), or (ii) within [***] days after Biogen's receipt of the Trigger Notice in all other cases (each, the "***Notice Review Period***"). As soon as reasonably practicable but not longer than the later of [***] days after Biogen's receipt of the Trigger Notice or [***] Business Days after Kiniksa's receipt of a Negotiation Notice, Kiniksa will provide Biogen with confidential materials and data with respect to (1) the Product and (2) any other products and programs that are part of the ROFN Transaction if the ROFN Transaction is with respect to (A) a Change of Control Transaction involving an Unsolicited Offer or (B) the Product together with any products or programs other than the Product,; provided, that, solely to the extent that (y) the ROFN Transaction is with respect to a Change of Control Transaction involving an Unsolicited Offer and (z) [***] Kiniksa will not be obligated under this Section 3.4.1(b) to provide Biogen with any other information regarding any other products and programs of Kiniksa (other than the Product) that are [***] Kiniksa determines in good faith, after consultation with outside counsel, that such action would be inconsistent with its fiduciary duties to the stockholders of Kiniksa under applicable law, which shall include in any case (1) an update of the information previously provided by Kiniksa in accordance with Section 3.3.2 for the Product (and, subject to the limitation in subsection (b)(2) above, any other information, products and programs that are the subject of or part of the ROFN Transaction), (2) to the extent not included as part of the foregoing update, any material clinical data and preclinical data with respect to the Product (and, subject to the limitation in subsection (b)(2) above, any other information, products and programs that are the subject of or part of the ROFN Transaction) Controlled by Kiniksa (each, a "***Data Package***"), and (3) such other information relating to the foregoing in Kiniksa's Control that Biogen may reasonably request at any time during the Diligence Period, which Data Package and additional requested information provided by Kiniksa to Biogen shall be Confidential Information of Kiniksa for purposes of this Agreement.

(c) During the period commencing on the date of receipt by Biogen of a Data Package and continuing for a period of (i) [***] days in the case where the Data Package is delivered with respect to a Change of Control Transaction involving an Unsolicited Offer, or (ii) [***] days in all other cases (each, the "***Diligence Period***"), Biogen will complete its diligence and Biogen and Kiniksa shall have periodic meetings (either in person or by phone) to discuss Biogen's progress and to answer any questions related to diligence.

(d) During the period commencing on the date of receipt by Biogen of a Data Package and continuing for a period of (i) [***] days in the case where the Data Package is delivered with respect to a Change of Control Transaction involving an Unsolicited Offer, or (ii) [***] days in all other cases (each, the "***Exclusivity Term***", which Exclusivity Term shall include the Diligence Period), Kiniksa will [***]; provided, that, to the extent that (i) [***] and (ii) the proposed ROFN Transaction is with respect to a Change of Control Transaction involving an Unsolicited Offer, Kiniksa shall be entitled during the Exclusivity Term to [***]

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to any Third Party if the Board of Directors or similar governing body of Kiniksa determines in good faith, after consultation with outside counsel, that the failure to take such action would be inconsistent with its fiduciary duties to the stockholders of Kiniksa under applicable law. During the Exclusivity Term, Kiniksa will exclusively negotiate in good faith with Biogen for, and Biogen will negotiate in good faith with Kiniksa for (A) in the case where the Data Package is with respect to a Change of Control Transaction involving an Unsolicited Offer, a term sheet, if accepted by each Party in its sole discretion, for an exclusive license or asset purchase of the Product or a Change of Control Transaction by and between the Parties, which term sheet shall include an extension of the Exclusivity Term sufficient for the Parties to negotiate and finalize an agreement with respect to the transaction and other terms acceptable to Kiniksa, in its sole discretion, or (B) in all other cases, an exclusive license or asset purchase agreement for the Product, or acquisition agreement with respect to Kiniksa, in each case on terms that are acceptable to each Party in its sole discretion; provided, that, Kiniksa shall be entitled to negotiate with Third Parties with respect to a Change of Control Transaction involving an Unsolicited Offer during the Exclusivity Term if the Board of Directors or similar governing body of Kiniksa determines in good faith, after consultation with outside counsel, that a failure to take such action would be inconsistent with its fiduciary duties to the stockholders of Kiniksa under applicable law. For the avoidance of doubt, each Party shall be entitled to reject any and all proposals made by the other Party during any Exclusivity Term in its sole discretion, without penalty. Notwithstanding anything to the contrary, nothing in this Section 3.4 shall be deemed to prevent Biogen from making an offer, solicited or unsolicited, at any time relative to an acquisition of Kiniksa.

(e) If (i) Biogen (A) does not deliver a Negotiation Notice to Kiniksa within the applicable Notice Review Period, or (B) fails to notify Kiniksa in writing that it wants to pursue an ROFN Transaction with respect to the Product or Change of Control Transaction, as the case may be, after review of the Data Package by the end of the applicable Diligence Period, or (ii) Biogen and Kiniksa do not mutually agree on the terms of a transaction (or a term sheet for the same, as applicable) within the Exclusivity Term, Kiniksa will be free to negotiate an ROFN Transaction for such Product, which may include a Change of Control Transaction, with any Third Party. Notwithstanding the prior sentence, if Kiniksa does not enter into a definitive agreement for a ROFN Transaction with respect to the Product or a Change of Control Transaction with any Third Party on or before [***] days from the date of expiration of the applicable period in subsections (i)(A), (i)(B) or (ii) above (or, if such definitive agreement is entered into within that period, such transaction is not consummated on or before [***] months after the expiration of such [***] period or is not consummated on or before [***] months after the expiration of such one [***] period solely to the extent the ROFN Transaction is with respect to a Change of Control Transaction with a Third Party that has [***]), Kiniksa shall provide Biogen with written notice and Biogen's Right of First Negotiation in this Section 3.4 shall immediately apply once again to such Product; provided, that, Kiniksa may [***] period during which Kiniksa has the right to enter into a definitive agreement for such ROFN Transaction (including a Change of Control Transaction) by an additional [***] days by providing written notice to Biogen [***] on or before the expiration of such [***] day period.

3.4.2 Definition of Change of Control Transaction. For the purposes of this Section 3.4, a "***Change of Control Transaction***" means (a) a transaction or series of related transactions pursuant to which any Third Party or group of related Third Parties (e.g., two or more Third Parties that act as a group for purposes of Section 13(d) of the Securities Exchange Act of 1934 (the "***Exchange Act***")) directly or indirectly would first become the beneficial owner of capital stock representing fifty percent (50%) or more of the total voting rights of Kiniksa and/or an Affiliate then outstanding or would first acquire the power to elect or appoint fifty percent (50%) or more of the members of the Board of Directors of Kiniksa and/or an Affiliate, (b) a transaction in which Kiniksa and/or an Affiliate proposes to sell to a Third Party all or substantially all of its assets, or (c) a merger transaction with a Third Party in which the stockholders of record of Kiniksa and/or an Affiliate immediately prior to the consummation of such transaction would not beneficially own capital stock representing fifty percent (50%) or more of the total voting rights of the surviving entity after the consummation of such transaction.

3.4.3 Definition of Exempt Transaction. For the purposes of this Section 3.4, an "***Exempt Transaction***" means any transaction by and between Kiniksa and (a) any Third Party engaged by Kiniksa to perform designated research or development activities (including drug development and/or manufacturing services) with respect to the Product, including any services or sponsored research agreement with a contract research organization, a contract manufacturing organization and/or university or other non-profit institution, or (b) any Third Party appointed by Kiniksa or any of its Affiliates (including a contract sales organization) to distribute, market or sell any Product, where such Third Party purchases its requirements of such Product from Kiniksa or its Affiliates for a transfer price but does not make any royalty, profit share or similar payment to Kiniksa based on sales of such Product.

3.4.4 Termination of Right of First Negotiation. Biogen's Right of First Negotiation described in this Section 3.4 will terminate and be of no further effect on the earliest of (a) the date upon which Kiniksa has a first commercial sale of the Product in the United States; (b) the [***] year anniversary of the Effective Date; or (c) with respect to a Change of Control Transaction, upon the consummation of a *bonafide* Change of Control Transaction between Kiniksa and a Third Party that has [***] after application of any of subsections 3.4.1(e)(i)(A), (e)(i)(B) or (e)(ii), to the extent applicable, and within the time periods specified in subsection 3.4.1(e).

4. PROSECUTION/INFRINGEMENT OF ACQUIRED PATENTS

4.1 **Prosecution of Acquired Patents**.

4.1.1 Responsibilities of Kiniksa. Kiniksa, acting through patent counsel or agents of its choice, shall be solely responsible for the preparation, filing, prosecution and maintenance of all Acquired Patent Rights throughout the Territory. All patent costs and expenses incurred by Kiniksa in connection with the preparation, filing, prosecution and maintenance of such Acquired Patent Rights shall be the sole responsibility of Kiniksa. Kiniksa hereby acknowledges its duties, including the duty of disclosure under 37 CFR 1.56 with respect

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to the Acquired Patent Rights.

4.1.2 Patent Term Extensions. Kiniksa shall use reasonable efforts to obtain all patent term extensions or supplemental protection certificates or their equivalents in any country where applicable to the Acquired Patent Rights. Biogen shall cooperate with Kiniksa with respect to such matters, including by timely conferring with Kiniksa to ensure compliance with applicable filing deadlines, and conferring with Kiniksa on the procedures to be followed by Kiniksa to ensure such compliance.

4.2 **Infringement**.

4.2.1 Notice. If either Party becomes aware of (a) any suspected infringement or misappropriation of any Acquired Patent Rights or (b) the submission by any Third Party of an abbreviated BLA under the Biologics Price Competition and Innovation Act for any Product (each, an "***Infringement***"), that Party shall promptly notify the other Party and provide it with all details of such Infringement of which it is aware (each, an "***Infringement Notice***").

4.2.2 Kiniksa Right. Kiniksa shall have the first right, but not the obligation, to address such Infringement in the Territory that involves such Acquired Patent Rights by taking reasonable steps, which may include the initiation of legal proceedings or actions to persuade the infringer to desist, compromise or otherwise settle such Infringement (each, an "***Infringement Response***"); provided, that: (a) Kiniksa shall keep Biogen informed about such Infringement Response and Biogen shall provide all reasonable cooperation to Kiniksa in connection with such Infringement Response; and (b) Kiniksa shall not take any position with respect to, or compromise or settle, any such Infringement in any way without first providing Biogen with (i) notice of Kiniksa's preferred course of action and (ii) an opportunity to provide comments, which comments Biogen will provide promptly and in any event within seven (7) days from receipt of such notice from Kiniksa and which comments Kiniksa will consider in good faith; and (c) if Kiniksa does not intend to prosecute or defend an Infringement with respect to any Acquired Patent Rights, or ceases to diligently pursue an Infringement Response with respect to such an Infringement, it shall inform Biogen in such a manner that such Infringement Response will not be prejudiced and Section 4.2.3 shall apply. All costs, including, without limitation, attorneys' fees, relating to such Infringement Response shall be borne solely by Kiniksa.

4.2.3 Biogen Right. If (a) Kiniksa informs Biogen that it does not intend to pursue any Infringement Response with respect to any Acquired Patent Rights, (b) within [***] days after the receipt of notice of any such Infringement, Kiniksa has not commenced to take any Infringement Response with respect thereto, or (c) if Kiniksa ceases to reasonably pursue any such Infringement Response, then Biogen shall have the right, but not the obligation, to

direct Kiniksa to take appropriate action to address such Infringement, including by instructing Kiniksa to initiate an Infringement Response or to continue prosecution of any legal proceedings initiated by Kiniksa; provided, that, Biogen shall first provide Kiniksa with a reasonable opportunity to explain the rationale for not pursuing or continuing such Infringement Response,

which Biogen will consider in good faith. In the event Biogen directs Kiniksa to pursue an Infringement Response, Kiniksa shall (i) to the extent it is not prohibited from doing so, join Biogen as a plaintiff in such Infringement action, (ii) keep Biogen fully informed about such Infringement Response and (iii) take any actions that may be reasonably requested by Biogen with respect to that Infringement Response. Kiniksa shall not take any position with respect to, or compromise or settle, any such Infringement in any way other than as directed by Biogen in writing. All costs, including, without limitation, attorneys' fees, relating to such Infringement Response shall be borne solely by Biogen. Notwithstanding the foregoing, Biogen may not compel Kiniksa to pursue an Infringement Response if Kiniksa reasonably determines that (a) such action would reasonably be expected to result in an action or proceeding seeking to invalidate any Acquired Patent Rights or declare any of the Acquired Patent Rights unenforceable or not infringed, and (b) the ability of Kiniksa to prevail in such invalidity, unenforceability or non-infringement action is uncertain.

4.2.4 Cooperation. In any Infringement Response instituted under this Section 4.2, the Parties shall cooperate with and assist each other in all reasonable respects.

4.2.5 Allocation of Recoveries. Any settlements, damages or monetary awards (collectively, a "***Recovery***") recovered by either Party pursuant to any Infringement Response shall, after reimbursing the Parties for their reasonable out-of-pocket expenses in making such recovery (which amounts shall be allocated pro rata if insufficient to cover the totality of such expenses) be allocated as follows: (a) to the extent such Infringement Response is instituted by Kiniksa, [***]; provided, that, [***]; and (b) [***].

5. PURCHASE PRICE AND OTHER PAYMENTS

5.1 **Purchase Price**.

5.1.1 Upfront Fee. Within thirty (30) days of the Effective Date, Kiniksa shall pay Biogen a one-time, non-refundable, non-creditable upfront fee in the amount of Eleven Million Five Hundred Thousand Dollars ($11,500,000) (the "***Upfront Fee***"), payable by electronic funds transfer of immediately available funds to an account or accounts specified to Kiniksa by Biogen in writing at least five (5) Business Days prior to the date thereof.

5.1.2 Technology Transfer Fee. Within thirty (30) days of the Completion of Technology Transfer, Kiniksa shall pay Biogen a one-time, non-refundable, non-creditable technology transfer fee in the amount of Five Hundred Thousand Dollars ($500,000) (the "***Technology Transfer Fee***"), payable by electronic funds transfer of immediately available funds to an account or accounts specified to Kiniksa by Biogen in writing at least five (5) Business Days prior to the date thereof.

5.2 **Milestone Payments**.

5.2.1 Development, Regulatory and Commercialization Milestones. Kiniksa

shall make the following one-time, non-refundable, non-creditable payments to Biogen within forty-five (45) days after the first achievement of each of the following milestone events by Kiniksa, its Affiliates, Distributors and/or licensees with respect to each Product that achieves each such milestone:

Milestone Event		Milestone Payment ($ U.S. Dollars)
Initiation of Phase I Clinical Trial with a Product	$	4,000,000
[***]	$	[***]

Milestone Event		Milestone Payment ($U.S. Dollars)
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]

[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]

[***]	$	[***]
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]

5.2.2 No Additional Milestone Payments. For purposes of clarity, (i) to the extent that any Product achieves the [***] milestone events [***], no additional milestone payments will be due and payable with respect to such Product pursuant to Section 5.2.1, (ii) to the extent that any Product achieves any [***] milestone event [***], no additional milestone payments will be due and payable with respect to such Product pursuant to Section 5.2.1, and (iii) [***]. For example, [***], then [***] and so on.

5.2.3 Sales Milestones. In addition to the milestone payments contemplated by Section 5.2.1, Kiniksa shall make each of the following one-time, non-refundable, non-creditable payments to Biogen within forty-five (45) days after the first occurrence of the corresponding milestone event by Kiniksa, its Affiliates and/or licensees with respect to any Product:

Milestone Event		Milestone Payment
[***]	$	[***]
[***]	$	[***]
[***]	$	[***]

5.2.4 Notice and Payment of Milestones. Kiniksa shall provide Biogen with prompt written notice upon the occurrence of each milestone event set forth in Section 5.2.1 and/or Section 5.2.3. If Biogen believes any such milestone event has occurred and has not received a written notice of same from Kiniksa, it shall so notify Kiniksa in writing and shall provide to Kiniksa documentation or other information that supports its belief. Any dispute under this Section 5.2.4 that relates to whether or not a milestone event has occurred shall be referred to both Party's executive officers by either Party, and, to the extent not resolved by the executive officers within thirty (30) days, shall be resolved in accordance with Section 10.3.

5.3 **Payment of Royalties; Royalty Rates**.

5.3.1 Payment of Royalties. Subject to Section 5.3.2, Kiniksa shall pay Biogen a royalty on Annual Net Sales of each Product by Kiniksa, its Affiliates, and licensees in each Calendar Year (or partial Calendar Year), commencing with the First Commercial Sale of such Product in any country in the Territory and ending upon the last day of the Royalty Term for such Product in such country, at the following rates:

Annual Net Sales Increment	Royalty Rate (%)
Annual portion less than [***]	[***]
Annual portion equal to or greater than [***]	[***]
Annual portion equal to or greater than [***]	[***]

For purposes of clarity, (i) each royalty rate will only apply to the corresponding tier of annual Net Sales, and (ii) the determination of the royalty rate under this Section 5.3.1 shall be based on aggregate, worldwide Annual Net Sales in each Calendar Year rather than on a country-by- country basis.

5.3.2 Adjustments to Royalties.

(a) No Patent Coverage. Notwithstanding anything to the contrary in Section 5.3.1, if any Product is sold by Kiniksa in a country and is not Covered by a Valid Claim of [***] in such country and such Product does not otherwise have regulatory exclusivity in such country, the royalty rates in such country shall be reduced by [***] of the rates set forth in Section 5.3.1, continuing until the last day of the applicable Royalty Term with respect to such Product in such country. The Parties hereby acknowledge and agree that royalties that are payable for a Product for which no [***] exist shall be in consideration of (i) Biogen's expertise and know-how concerning its development of the Acquired Know-How prior to the Effective Date; (ii) the transfer to Kiniksa hereunder of Acquired Know-How that is not within the claims of any Patent Rights Controlled by Biogen; and (iii) the "head start" afforded to Kiniksa by each of the foregoing.

(b) Royalty Stacking. The amount of royalties payable to Biogen under Section 5.3.1 for any Product in any country shall be reduced by [***] paid by Kiniksa or

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

any of its Affiliates to any Third Party in consideration for the license of [***] the manufacture, use or sale of the Product in such country in the absence of such a license; provided, that, in no event shall the royalty payments owed under Section 5.3.1 with respect to a Product in a country be reduced by operation of this Section 5.3.2(b) by more than [***] of what would otherwise be owed under Section 5.3.1; provided, further, that, (i) Kiniksa will be entitled to carry forward and apply against royalty payments due and payable in subsequent Calendar Years any amounts with respect to which Kiniksa would have been entitled to make a deduction pursuant to this Section 5.3.2(b) but for such maximum annual reduction and (ii) the right of Kiniksa to carry forward any amounts pursuant to this Section 5.3.2(b) will expire upon the expiration of the Term.

(c) Competing Drugs. In the event that one or more Third Parties (other than an Affiliate or licensee of Kiniksa) sell a Competing Drug (as defined below) in any country in which a Product is then being sold by Kiniksa then, (i) during any Calendar Quarter in which sales of the Competing Drug by such Third Parties are equal to or greater than [***] of aggregate unit sales of Products and Competing Drugs in such country (as measured by prescriptions or other similar information available from a Third Party data provider and applicable to such country) the applicable royalties in effect with respect to such Product in such country as specified in Section 5.3.1 shall be reduced by [***] and (ii) during any Calendar Quarter in which sales of the Competing Drug by such Third Parties are equal to or greater than [***] of aggregate unit sales of Products and Competing Drugs in such country (as measured by prescriptions or other similar information available from a Third Party data provider and applicable to such country) the applicable royalties in effect with respect to such Product in such country as specified in Section 5.3.1 shall be reduced by [***]. Notwithstanding the foregoing, (a) Kiniksa's obligation to pay royalties at [***] of the applicable royalty rates shall be reinstated on the first day of the Calendar Quarter immediately following the Calendar Quarter in which sales of such Competing Drugs account for less than [***] but equal to or greater than [***] of aggregate unit sales of Products and Competing Drugs in such country and (b) Kiniksa's obligation to pay royalties at the full royalty rates shall be reinstated on the first day of the Calendar Quarter immediately following the Calendar Quarter in which sales of such Competing Drugs account for less than [***] of aggregate unit sales of Products and Competing Drugs in such country. For purposes of this Section 5.3.2(c), (a) a "***Competing Drug***" means, with respect to a Product, a therapeutic product that (i) [***], (ii) [***] and (iii) [***] and (b) "***Bioequivalent***" or "***Bioequivalence***" means, a biological product that (i) is highly similar to the Product notwithstanding minor differences in clinically inactive components; and (ii) has no clinically meaningful differences between the biological product and the Product in terms of the safety, purity, and potency.

5.3.3 Maximum Reduction Amount. Notwithstanding anything to the contrary in this Section 5.3.3, in no event will the reductions in Section 5.3.2(a) and/or Section 5.3.2(b) and/or Section 5.3.2(c) reduce the royalty rates under Section 5.3.1 in any Calendar Year to less than [***] of the royalty rates set forth in Section 5.3.1.

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

5.3.4 Payment Dates and Reports. Royalty payments shall be made by Kiniksa with respect to each Product within forty-five (45) days after the end of each Calendar Quarter in which a sale of such Product is made, commencing with the Calendar Quarter in which the First Commercial Sale of such Product occurs. Kiniksa shall also provide, at the same time each such payment is made, a report showing: (a) the Net Sales of each Product by type of Product and country in the Territory; (b) the total amount of deductions from Gross Sales to determine Net Sales; (c) the applicable royalty rates for each Product on a country-by-country basis in each country in the Territory after applying any adjustments set forth in Section 5.3.2 above; (d) a calculation of the amount of royalty due to Biogen; and (e) the expected date of expiration of regulatory exclusivity of each Product in each country in the Territory where such Product is being sold.

5.3.5 Records; Audit Rights. Kiniksa and its Affiliates and licensees shall keep and maintain for [***] years from the date of each payment of royalties hereunder complete and accurate records of Gross Sales and Net Sales by Kiniksa and its Affiliates and licensees in sufficient detail to allow royalties to be determined accurately. Biogen shall have the right for a period of [***] years after receiving any such payment to appoint at its expense an independent certified public accountant reasonably acceptable to Kiniksa to audit the relevant records of Kiniksa and its Affiliates and licensees to verify that the amount of such payment was correctly determined. Kiniksa and its Affiliates and licensees shall each make its records available for audit by such independent certified public accountant during regular business hours at such place or places where such records are customarily kept, upon thirty (30) days written notice from Biogen. Such audit right shall not be exercised by Biogen more than once in any Calendar Year or more than once with respect to sales of a particular Product in a particular period. All records made available for audit shall be deemed to be Confidential Information of Kiniksa. The results of each audit, if any, shall be binding on both Parties absent manifest error. In the event there was an underpayment by Kiniksa hereunder, Kiniksa shall promptly (but in any event no later than thirty (30) days after Kiniksa's receipt of the report so concluding) make payment to Biogen of any shortfall. Biogen shall bear the full cost of such audit unless such audit discloses an underreporting by Kiniksa of [***] percent ([***]%) of the aggregate amount of royalties payable in any Calendar Year, in which case Kiniksa shall reimburse Biogen for all costs incurred by Biogen in connection with such audit.

5.4 **Payments Under Retained Contracts**. Kiniksa hereby acknowledges that Biogen is obligated to make payments owed to certain Third Parties under the Retained Contracts on and after the Effective Date. Kiniksa shall be responsible for making payments to Biogen for the amount of such payments, including a portion of the annual maintenance fees that are applicable to the Development and/or Commercialization by Kiniksa of Acquired Antibodies and/or Products as set forth on Schedule E ("***Retained Contract Payments***"). Kiniksa shall make such Retained Contract Payments directly to Biogen, and in each such instance, Kiniksa shall make the requisite payments to Biogen and provide the necessary reporting information to Biogen in sufficient time to enable Biogen to comply with its obligations under such Retained Contracts. Kiniksa shall be entitled to credit up to [***] of any

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

and all such Retained Contract Payments against royalty payments due and payable by Kiniksa to Biogen under Section 5.3.

5.5 **Payments in Dollars**. All payments made by Kiniksa under this Article 5 shall be made by wire transfer from a banking institution in United States Dollars in accordance with instructions given by Biogen in writing to Kiniksa from time to time.

5.6 **Foreign Currency Exchange**. If, in any Calendar Quarter, Net Sales are made in any currency other than United States Dollars, such Net Sales shall be converted into United States Dollars as follows:

(A/B), where

A = foreign "Net Sales" (as defined above) in such Calendar Quarter expressed in such foreign currency; and

B = the applicable foreign exchange conversion rate, expressed in local currency of the foreign country per United States Dollars (using, as the applicable foreign exchange rate, the average of the daily closing rates published in *Bloomberg* or any other mutually agreed upon source, for such Calendar Quarter).

5.7 **Overdue Payments**. All undisputed payments not made by Kiniksa to Biogen when due under this Agreement shall [***] from the due date until paid in full or, if less, the maximum interest rate permitted by applicable Law. Any such overdue payment shall, when made, be accompanied by, and credited first to, all interest so accrued. If Kiniksa has a good faith dispute regarding a payment to be made to Biogen, Kiniksa may withhold payment for the disputed amount; provided; that, Kiniksa pays all undisputed amounts and notifies Biogen in writing of the specific amount and nature of the dispute on or before the due date for the payment.

5.8 **Taxes**.

5.8.1 Payments. The payments set forth in Article 5 shall not be reduced by any Transfer Taxes which, if charged, shall be payable by Kiniksa pursuant to Section 5.8.2.

5.8.2 Transfer Taxes. All transfer, documentary, sales, use, valued-added, gross receipts, stamp, registration or other similar transfer Taxes incurred in connection with the transfer and sale of the Purchased Assets or the license of the Acquired Know-How as contemplated by the terms of this Agreement ("***Transfer Taxes***"), including all recording or filing fees, notarial fees and other similar costs, that may be imposed, payable, collectible or incurred shall be borne by Kiniksa. The Parties have determined that the value of the tangible property transferred pursuant to Section 2.1 is [***] of the payment made pursuant to Section 5.1 is allocated to consideration for such tangible property. Each Party shall cooperate and provide such assistance to the other Party, including the provision of such documentation as may be required by a tax authority or other Governmental Entity, as may be reasonably necessary in

order to reduce or eliminate the amount of any Transfer Taxes described in the first sentence of this Section 5.8.2 in a manner consistent with applicable Laws.

5.8.3 Tax Withholding. Kiniksa and its Affiliates shall be entitled to deduct and withhold any Taxes, withholding or similar amount required by applicable Law to be deducted and withheld (other than with respect to any Transfer Taxes for which Kiniksa is responsible pursuant to Section 5.8.2) with respect to any amount payable to Biogen, any of its Affiliates or any Person described in clause (b) of Section 10.6 in connection with the transactions and/or payments contemplated by this Agreement. Kiniksa shall use commercially reasonable efforts to notify Biogen in writing of such withholding requirements prior to making the payment to Biogen and to provide such assistance to Biogen, including the provision of such documentation as may be required by a Tax authority or other Governmental Entity, as may be reasonably necessary in Biogen's efforts to claim an exemption from or reduction of such Taxes. Kiniksa will, in accordance with Law, withhold Taxes from the amount due, remit such Taxes to the appropriate tax authority or other Governmental Entity, and furnish Biogen with proof of payment of such Taxes within fifteen (15) days following payment thereof. If Taxes are paid to a tax authority or other Governmental Entity, Kiniksa shall use commercially reasonable efforts to provide such assistance to Biogen (at Biogen's expense) as is reasonably required to obtain a refund of Taxes withheld, or obtain a credit with respect to Taxes paid. If [***] Kiniksa will [***] *minus* (i) [***]; provided, however, that Kiniksa shall not be required to [***] but for [***].

6. REPRESENTATIONS AND WARRANTIES

6.1 Mutual Warranties. Each of Kiniksa and Biogen represents and warrants to the other Party that:

(a) it is duly organized and validly existing under the Law of the jurisdiction of its incorporation, and has full corporate power and authority to enter into this Agreement and to carry out the provisions hereof;

(b) it is duly authorized to execute and deliver this Agreement and to perform its obligations hereunder, and the individual executing this Agreement on its behalf has been duly authorized to do so by all requisite corporate action; and

(c) this Agreement is legally binding upon it and enforceable in accordance with its terms. The execution, delivery and performance of this Agreement by it does not conflict with any agreement, instrument or understanding, oral or written, to which it is a party or by which it may be bound, nor violate any material applicable Law.

6.2 Additional Biogen Warranties. Biogen represents and warrants to Kiniksa that, as of the Effective Date**:**

6.2.1 Title to Assets. Biogen or its Affiliates have good and valid title to all of

the Purchased Assets in their entirety (subject to the grant back license to Biogen in Section 2.5.2) free and clear of all Encumbrances, except for Permitted Encumbrances.

6.2.2 No Debarment. Neither Biogen nor its Affiliates' employees who have been involved in the Exploitation of BIIB069, nor, to Biogen's Knowledge, any employees of their respective licensees, contractors, agents and consultants who have been involved, on behalf of Biogen, in the Exploitation of BIIB069:

(a) is debarred under Section 306(a) or 306(b) of the FD&C Act or by the analogous applicable Laws of any Regulatory Authority;

(b) has been charged with, or convicted of, any felony or misdemeanor within the ambit of 42 U.S.C. §§ 1320a-7(a), 1320a-7(b)(l)-(3), or pursuant to the analogous applicable Laws of any Regulatory Authority, or is proposed for exclusion, or the subject of exclusion or debarment proceedings by a Regulatory Authority; or

(c) is excluded, suspended or debarred from participation, or otherwise ineligible to participate, in any U.S. or non-U.S. healthcare programs (or has been convicted of a criminal offense that falls within the scope of 42 U.S.C. §1320a-7 but not yet excluded, debarred, suspended, or otherwise declared ineligible), or excluded, suspended or debarred by a Regulatory Authority from participation, or otherwise ineligible to participate, in any procurement or non- procurement programs.

6.2.3 Litigation and Claims. There is no action, suit, claim, proceeding or investigation pending that has been served on Biogen, and to the Knowledge of Biogen, there is no other action, suit, claim, proceeding or investigation pending or threatened against Biogen before or by any federal, state, municipal or other governmental court, agency or instrumentality, which would prevent Biogen's performance of this Agreement and the transactions contemplated hereby.

6.2.4 Intellectual Property Rights.

(a) Biogen has sufficient legal and/or beneficial ownership and/or rights in the Acquired Patent Rights and Acquired Know-How necessary to assign and transfer to Kiniksa the Acquired Patent Rights and Acquired Know-How in accordance with the terms of this Agreement.

(b) None of the Acquired Patent Rights or Acquired Know-How constitute Third Party Intellectual Property.

(c) The Acquired Patent Rights have been duly filed in the jurisdictions identified in Part 1 of Schedule A, are pending, have not been abandoned or allowed to lapse, and have not been held invalid or unenforceable by a decision of a court or other governmental agency of competent jurisdiction, in whole or in part, nor to Biogen's

Knowledge, is there any reason for the Acquired Patent Rights to be deemed invalid or held unenforceable by a decision of a court or other governmental agency of competent jurisdiction.

(d) To the Knowledge of Biogen, there are no oppositions, cancellations, interferences or Litigation proceedings pending or expressly threatened in writing, challenging the ownership, validity or enforceability of any of the Acquired Patent Rights, or, to the Knowledge of Biogen, any of the Background Licensed Patent Rights.

(e) Part 1 of Schedule A accurately sets forth for each provisional or pending patent application in the Acquired Patent Rights, the application number, date of filing and title for each country, and listing, as applicable, deadlines for any renewals or other required filings or payments within ninety (90) days after the Effective Date.

(f) Neither Biogen, nor any of Biogen's Affiliates, has received from any Person, any actual or, to the Knowledge of Biogen, threatened claim that the use of the Acquired Patent Rights, Acquired Know-How, Background Licensed Patent Rights, Background Sublicensed Patent Rights, as has been and is now being conducted, presently infringes or constitutes a misappropriation of any registered patents of any Person. Biogen has not granted any licenses or covenants not to sue under the Acquired Patent Rights, except under the Amgen Agreement. Biogen has paid all licensing fees, royalties, profit participations and other payments that were due or payable by Biogen or any of its Affiliates in connection with its use or practice of the Acquired Patent Rights, Acquired Know-How and Background Licensed Patent Rights prior to the Effective Date. Biogen has paid all licensing fees, royalties, profit participations and other payments that were due or payable by Biogen or any of its Affiliates through the Effective Date in connection with its use or practice of the Background Sublicensed Patent Rights prior to the Effective Date.

(g) Except as described on Schedule I attached hereto, (i) the list of Acquired Patent Rights included on Part 1 of Schedule A and, (ii) to the Knowledge of Biogen, the list of Acquired Know-How included on Part 2 of Schedule A attached hereto, the list of Background Licensed Patent Rights included on Schedule D attached hereto, and the Background Sublicensed Intellectual Property in-licensed by Biogen pursuant to the Retained

Contracts is a complete and accurate list of all Know-How and Patent Rights owned or Controlled by Biogen or its Affiliates prior to the Effective Date that were used by Biogen in, and necessary for, the Exploitation of the Acquired Antibody as it exists as of the Effective Date. To Biogen's Knowledge, neither Biogen nor its Affiliates own or Control any intellectual property rights other than the intellectual property rights set forth in the list of Acquired Patent Rights and Acquired Know-How included on Parts 1 and 2 of Schedule A attached hereto, the list of Background Licensed Patent Rights included on Schedule D attached hereto, and the Background Sublicensed Intellectual Property in-licensed by Biogen pursuant to the Retained Contracts that were used by Biogen in, and necessary for, the Exploitation of the Acquired Antibody as it exists as of the Effective Date.

(h) Biogen is the sole and exclusive owner of, or Controls, the

Background Licensed Patent Rights included on Schedule D attached hereto

(i) Biogen has the ability to grant to Kiniksa the licenses or sublicenses to the Background Licensed Patent Rights and Background Sublicensed Patent Rights, as the case may be, granted to Kiniksa under this Agreement.

(j) To Biogen's Knowledge, Biogen has complied in all material respects with all applicable Laws in connection with the prosecution and maintenance of the Acquired Patent Rights claiming any Acquired Antibody or any aspect of the Exploitation thereof, in the Field and in the Territory.

(k) To Biogen's Knowledge, Biogen has disclosed to the U.S. Patent and Trademark Office all information in Biogen's possession or control as of the Effective Date that is required to be disclosed under 37 C.F.R. § 1.56 for prosecuting the Acquired Patent Rights.

(l) Except as set forth on Schedule I attached hereto, Biogen is not directly or indirectly (i) conducting, participating in or sponsoring any activities that are directed toward the research, development, manufacture, sales, marketing, promotion or distribution of any compound or biologic that [***] or (ii) seeking to appoint, grant any right or license to or otherwise authorize any Third Party to perform any of the foregoing, or (iii) actively seeking to acquire any right or license to any compound or biologic that [***].

(m) Biogen has not received written notice that it has materially breached its obligations under the Assigned Contract in a manner that has, or would reasonably be expected to have, a material adverse effect on the rights granted to Kiniksa under this Agreement.

(n) Biogen has not received written notice that it has materially breached its obligations under any of the Retained Contracts in a manner that has, or would reasonably be expected to have, a material adverse effect on the rights granted to Kiniksa under this Agreement.

6.3 **No Other Warranties**. EXCEPT FOR THE REPRESENTATIONS AND WARRANTIES EXPRESSLY SET FORTH IN THIS ARTICLE VI, BOTH PARTIES DISCLAIM ALL OTHER REPRESENTATIONS AND WARRANTIES, EXPRESS OR IMPLIED, STATUTORY OR OTHERWISE, WITH REGARD TO THE PURCHASED ASSETS AND THIS AGREEMENT, INCLUDING WARRANTIES OF MERCHANTABILITY, FITNESS FOR A PARTICULAR PURPOSE, VALIDITY AND NON- INFRINGEMENT OF INTELLECTUAL PROPERTY RIGHTS.

7. CONFIDENTIALITY

7.1 **Confidential Information**. The term "***Confidential Information***" shall mean

(a) the Acquired Know-How, (b) the Acquired Patent Rights, (c) information provided by Kiniksa under any Report, and (d) information provided by Kiniksa pursuant to Section 3.4; except that with respect to (c) or (d), such information shall not be considered Confidential Information to the extent that it can be established that such Confidential Information (i) was already known by Biogen at the time of disclosure to Biogen, (ii) was generally available to the public or otherwise part of the public domain at the time of its disclosure to Biogen, (iii) became generally available to the public or otherwise part of the public domain after its disclosure and other than through any act or omission of Biogen in breach of this Agreement, (iv) was disclosed to Biogen, by a Third Party whose disclosure does not, to Biogen's knowledge, constitute a breach of an obligation to Kiniksa, or (v) was subsequently developed by Biogen without the aid, use or application of the Confidential Information as demonstrated by competent written records. For purposes of clarity, on and after the Effective Date, all Confidential Information shall be considered the property of Kiniksa.

7.2 **Restrictions**. For a period of [***] years after the Effective Date, Biogen will keep all Confidential Information in confidence with the same degree of care with which Biogen holds its own confidential information but in no event with less than a reasonable degree of care. Notwithstanding anything to the contrary in the foregoing, Biogen's obligations of confidentiality and non-use with respect to any documents identified as "Extended Confidentiality" in Part 2 of Schedule A attached hereto shall survive for a period of [***] years after the Effective Date. Biogen will not use or disclose Confidential Information except in connection with the performance of its obligations under this Agreement. Biogen has the right to disclose Confidential Information without Kiniksa's prior written consent, to the extent and only to the extent reasonably necessary, to Biogen's Affiliates and their employees, subcontractors, consultants or agents who have a need to know such Confidential Information in order to perform its obligations under this Agreement and who are required to comply with the restrictions on use and disclosure in this Section 7.2; provided, that, Biogen assumes responsibility and remains liable for the compliance of such Affiliates and their employees, subcontractors, consultants and agents with such obligations. Biogen will use reasonable measures and precautions to cause those Persons to comply with the restrictions on use and disclosure in this Section 7.2.

7.3 **Exception**. Biogen's obligation of nondisclosure and the limitations upon the right to use the Confidential Information will not apply to the extent that Biogen can demonstrate that the Confidential Information is or becomes public knowledge through no act or omission of Biogen or any of its Affiliates. Disclosure of Confidential Information shall not be prohibited to the extent required to comply with applicable laws or regulations, or with a valid court or administrative order, provided that the Biogen: (a) promptly notifies the Kiniksa in writing of the existence, terms and circumstances of such required disclosure; (b) consults with the Kiniksa on the advisability of taking legally available steps to resist or narrow such disclosure; and (c) takes all reasonable and lawful actions to obtain confidential treatment for such disclosure.

7.4 **Publication by Biogen**. If Biogen wishes to make a publication or presentation

with respect to any Acquired Antibody or Product, Biogen shall (a) obtain the prior written consent of Kiniksa, which consent shall not be unreasonably withheld, conditioned or delayed;

(b) deliver to Kiniksa a copy of the proposed written publication or an outline of the proposed presentation at least thirty (30) days prior to submission for publication or presentation in order to give Kiniksa the opportunity to comment on such publication or presentation; and (c) consider all reasonable comments and proposed changes of Kiniksa to the proposed publication or presentation, provided, however, that Biogen shall not disclose any Confidential Information of Kiniksa in any such publication or presentation without Kiniksa's prior written consent.

7.5 **Terms of this Agreement; Publicity**.

7.5.1 Restrictions. The Parties agree that neither Party shall (i) disclose the existence or terms of this Agreement or the terms of any term sheet or agreement negotiated pursuant to Section 3.4, or (ii) issue any press release or public statement disclosing information relating to this Agreement or the transactions contemplated hereby or the terms hereof, or the terms of any term sheet or agreement negotiated pursuant to Section 3.4, without prior written consent of the other Party, which consent shall not be unreasonably withheld, conditioned or delayed (or as such consent may be obtained in accordance with Section 7.5.2). Notwithstanding the foregoing, either Party may disclose the existence and terms of this Agreement to its Affiliates, and to its (actual or potential) permitted licensees, sublicensees, acquirers or assignees and subcontractors (and their advisors) and to investment bankers, investors, lenders, accountants and legal advisors and to such Party's directors, employees, contractors and agents, who have a need to know such Confidential Information. Each Party shall advise any such permitted licensees, sublicensees, acquirers or assignees, subcontractors (and their advisors), investment bankers, investors, lenders, accountants and legal advisors and such Party's directors, employees, contractors and agents who receive Confidential Information of the confidentiality obligations set forth in Article 7, and such Party shall take steps to ensure (through enforcement of written agreements or otherwise) that they comply with such obligations as if they had been a Party hereto; provided, however, that such Party shall remain responsible for any failure by any Person who receives such information from such Party pursuant to this Section 7.5 to treat such information as required under this Article 7.

7.5.2 Review. In the event either Party (the "***Issuing Party***") is required by Law or the rules or regulations of any applicable United States securities exchange or regulatory or governmental body to which the relevant Party is subject to issue a press release or other public statement disclosing information relating to this Agreement or the transactions contemplated hereby or the terms hereof, the Issuing Party will provide the other Party (the "***Reviewing Party***") with a copy of the proposed press release or public statement (the "***Release***"). The Issuing Party will specify with each such Release, taking into account the urgency of the matter being disclosed, a reasonable period of time within which the Receiving Party may provide any comments on such Release (but in no event less than five (5) Business Days, unless earlier disclosure is required) and if the Receiving Party fails to provide any comments during the response period called for by the Issuing Party, the Reviewing Party will

be deemed to have consented to the issuance of such Release. If the Receiving Party provides any comments, the Parties will consult on such Release and work in good faith to prepare a mutually acceptable Release. Either Party may subsequently publicly disclose any information previously contained in any Release so consented to. For the avoidance of doubt, Kiniksa, in its sole discretion, may disclose the results or status of research, development or any Clinical Trial conducted by Kiniksa or any health or safety matter related to any Acquired Antibodies or Products.

8. TERM; TERMINATION; EFFECT OF TERMINATION

8.1 **Term**. This Agreement shall commence on the Effective Date and shall continue in full force and effect, unless otherwise terminated pursuant to Section 8.2, until the expiration of all payment obligations under this Agreement with respect to the last Product in all countries in the Territory (the "***Term***").

8.2 **Right to Terminate**.

8.2.1 Termination by Kiniksa. Kiniksa may terminate this Agreement, effective at any time, by providing not less than ninety (90) days' prior written notice to Biogen.

8.2.2 Termination by Mutual Consent. The Parties may terminate this Agreement, effective at any time, by mutual written consent.

8.2.3 Termination for Breach. If a Party materially breaches this Agreement, the non-breaching Party may provide the breaching Party with a written notice specifying the nature of the breach, and stating its intention to terminate this Agreement if such breach is not cured. Subject to Section 8.2.4, if the material breach is not cured within ninety (90) days (or thirty (30) days with respect to breach of a payment obligation) after the receipt of such notice, the non-breaching Party shall be entitled, without prejudice to any of its other rights under this Agreement, and in addition to any other remedies available to it by law or in equity, to terminate this Agreement by providing written notice to the other Party. The applicable cure period shall be tolled pending resolution of any bona fide dispute between the Parties as to whether any such material breach has occurred.

8.2.4 Termination of Rights of Kiniksa Under ATCC Agreement. If Kiniksa breaches any of its material obligations under the ATCC Agreement, Biogen may provide Kiniksa with a written notice specifying the nature of the breach, and stating its intention to terminate the sublicense under the ATCC Agreement granted to Kiniksa by Biogen under this Agreement if such breach is not cured. If the breach is not cured within ninety (90) days (or sixty (60) days for non-payment) after the receipt of such notice, Biogen shall be entitled, without prejudice to any of its other rights under this Agreement, and in addition to any other remedies available to it by law or in equity, to terminate the sublicense under the ATCC Agreement granted to Kiniksa under this Agreement by providing written notice to Kiniksa.

8.3 **Effect of Termination**. If this Agreement is terminated pursuant to Section 8.2, the following shall apply:

(a) the licenses and rights granted by Biogen to Kiniksa pursuant to Section 2.5.1 shall terminate;

(b) solely to the extent the effective date of termination is prior to the expiration of the Exclusivity Period, the obligations of Kiniksa in Section 2.7 shall survive and shall continue in full force and effect for a period of [***] months from the effective date of termination;

(c) Kiniksa shall, within [***] days of the effective date of such termination, take such actions and execute such documents, as may be reasonably required to re- assign to Biogen all of its right, title and interest in and to the Purchased Assets, including the Acquired Patent Rights and Acquired Know-How;

(d) upon the written request of Biogen, Kiniksa shall grant Biogen an exclusive, worldwide, perpetual, freely sublicensable license, under any Kiniksa Know-How and/or Kiniksa Patent Rights used in, and necessary for, the Exploitation of Acquired Antibodies and Products in the Territory as of the effective date of termination;

(e) to the extent that that Biogen provides the written request described in subsection (d), Kiniksa shall promptly (and in any event within [***] days, except as otherwise provided below) take the following actions and the following provisions shall apply:

(i) Kiniksa shall provide Biogen with a reasonably detailed summary, together with reasonable supporting documents, of the aggregate Development Costs incurred by Kiniksa with respect to each Acquired Antibody and/or Product through the effective date of termination;

(ii) Kiniksa shall (A) promptly provide Biogen with copies of any and all Kiniksa Third Party Agreements and (B) take such steps as may be reasonably required to assign such Kiniksa Third Party Agreements that relate solely to the Acquired Antibody and/or Product to Biogen (such agreements "***Assigned Kiniksa Agreements***"); provided, that, if any Kiniksa Third Party Agreement is not assignable or transferable pursuant to this subsection (ii) (such agreements, "***Non-Assignable Kiniksa Agreements***") then Kiniksa shall (1) continue to use commercially reasonable efforts to obtain consent to assign such Non- Assignable

Kiniksa Agreements to Biogen as soon as practicable after the effective date of termination and shall upon receipt thereof, promptly assign to Biogen such Non-Assignable Kiniksa Agreements, and (2) cooperate, and cause its Affiliates to cooperate, with Biogen in any reasonable arrangement designed to provide Biogen with all of the benefits of, subject to the related obligations under, such Non-Assignable Kiniksa Agreements as if the appropriate assignment had been obtained;

(iii) to the extent there are any Non-Assignable Kiniksa Agreements as provided in subsection (ii), (A) Biogen shall be responsible for (1) making any payments (including royalties, milestones and other amounts) incurred and payable by Kiniksa to any Third Parties after the effective date of termination under any such Non-Assignable Kiniksa Agreements that are applicable to the development and/or commercialization by Biogen of Acquired Antibodies and/or Products by making such payments directly to Kiniksa, and in each instance Biogen shall make the requisite payments to Kiniksa and provide the necessary reporting information to Kiniksa in sufficient time to enable Kiniksa to comply with its obligations under the Non-Assignable Kiniksa Agreements, and (2) complying with any other obligations included in the Kiniksa Non-Assignable Agreements that are applicable to the development and/or commercialization of Acquired Antibodies and/or Products; (B) Kiniksa shall be responsible for paying or providing to any such Third Party any payments or reports made or provided by Biogen under this Section 8.3(e)(iii) and will provide Biogen with written notice of its compliance with such obligations; (C) Kiniksa shall not [***] and will not undertake any action that would [***]; and (D) upon written notice to Kiniksa, Biogen, may, at any time and in its sole discretion, [***], upon which [***] for purposes of this Section 8.3(e)(iii) and Biogen shall have no further obligations to Kiniksa with respect to [***];

(iv) to the extent that at any time on and after the effective date of termination, Biogen, in its sole discretion, determines to develop and/or commercialize any Product (each, a "***Biogen Product***") (it being acknowledged by Kiniksa that Biogen shall have no obligation to develop or commercialize any Product after the effective date of termination and will not be subject to any diligence obligations, including the diligence obligations of Kiniksa in Section 3.2, in connection therewith), (A) Biogen shall pay Kiniksa an amount equal to the Applicable Multiplier times any milestone payments and

royalty payments that would otherwise be due and payable by Kiniksa for Products pursuant to Section 5.2 and Section 5.3 to the extent applicable to the development or commercialization of such Biogen Product by Biogen; (B) the terms set forth in Section 5.2 and Section 5.3, and all related obligations (including the right to offset and/or reduce payments in accordance with Section 5.3.2) shall apply *mutatis mutandis* to each such Biogen Product; (C) the obligation of Biogen to make any royalty payments with respect to any Biogen Product under this Section 8.3(e)(iv) shall terminate on [***]; (D) the obligation of Biogen to make any milestone payments with respect to any Biogen Product under this Section 8.3(e)(iv) shall terminate [***]; and (E) the [***] on and after the effective date of termination; provided, that, if the Agreement is terminated by Kiniksa pursuant to Section 8.2.3, then the obligation of Biogen to [***] on the date on which Biogen has [***];

(v) upon the written request of Biogen (which request may specify any or all of the actions in clauses (A) through (G) below), Kiniksa shall promptly (and in any event within [***] days after such request, except as otherwise provided below, or except as otherwise not possible within [***] days due to applicable regulatory procedures in a jurisdiction, in which case such actions shall be taken as promptly as reasonably possible): (A) at Biogen's election, execute an assignment to Biogen of, or a grant to Biogen of an exclusive, worldwide, license under, all Product Trademarks Controlled by Kiniksa and applicable to any Products solely for use in connection with the Commercialization of such Products in the Territory, if any, other than any such Product Trademarks that incorporate the Kiniksa name or logo, or any tagline used in connection with Kiniksa's business; (B) transfer to Biogen all of its right, title and interest in and to all Regulatory Filings and Marketing Authorizations then in its name applicable to any Acquired Antibodies and/or Products, if any, and all Confidential Information Controlled by it as of the date of termination relied on by such Regulatory Filings and Marketing Authorizations; (C) notify the applicable Regulatory Authorities and take any other action reasonably necessary to effect such transfer; (D) provide Biogen with copies of all correspondence between Kiniksa and such Regulatory Authorities relating to such Regulatory Filings and Marketing Authorizations; (E) unless expressly prohibited by any Regulatory Authority, transfer sponsorship and control to Biogen of all Clinical Trials of any Acquired Antibodies and/or Products being conducted by or on behalf of Kiniksa as of the effective date of termination and continue to conduct such Clinical

Trials after the effective date of termination to enable such transfer to be completed without interruption of any such trial for up to [***] months from the effective date of termination; (F) cooperate with Biogen, cause its Affiliates to cooperate with Biogen, and use Commercially Reasonable Efforts to require any Third Party with which Kiniksa has an agreement with respect to the conduct of Clinical Trials for any Acquired Antibodies and/or Products (including, without limitation, agreements with contract manufacturing organizations, contract research organizations, clinical sites and investigators) to cooperate with Biogen in order to accomplish the transfer to Biogen of rights to those held by Kiniksa under its agreements with such Third Parties as of the effective date of termination; and (G) provide Biogen with copies of all reports and data, including clinical data, generated or obtained by Biogen or its Affiliates pursuant to this Agreement that relate to any Acquired Antibodies and/or Products that have not previously been provided to Biogen;

(vi) upon the written request of Biogen, Kiniksa shall promptly, and in any event within [***] days after such request: (A) take such steps as may be reasonably necessary to assign to Biogen Kiniksa's rights under any supply agreement by and between Kiniksa and any Third Party manufacturer to the extent that it provides for the supply of any Acquired Antibodies and/or Products (each, a "**Kiniksa Supply Agreement**") to the extent permitted by the terms of the Kiniksa Supply Agreement; (B) consent to the supply by the Third Party manufacturer to Biogen of Biogen's requirements of such Acquired Antibodies and Products, to the extent permitted under the terms of the Kiniksa Supply Agreement; or (C) provide Biogen or its designee with reasonable assistance in order to facilitate (1) the transfer to Biogen of the manufacturing processes for such Acquired Antibodies and Products and any related manufacturing Know-How, in each case used by Kiniksa or such Kiniksa Third Party manufacturer with respect such Acquired Antibodies and Products, and (2) the qualification of Biogen's or its designee's facility as required by any Regulatory Authority in order for Biogen or its designee to manufacture quantities of such Acquired Antibodies and Products. Without limiting the generality of the foregoing, to the extent Biogen requests that Kiniksa undertake the steps in subsection (C) above, Kiniksa shall, and shall use Commercially Reasonably Efforts to cause any applicable Affiliate or Kiniksa Third Party manufacturer to:

(I) make available to Biogen or its designee all Know- How Controlled by Kiniksa and used by Kiniksa or such Kiniksa Third Party manufacturer with respect the manufacture of the Acquired Antibodies and Products, including documentation constituting material support, performance advice, shop practice, specifications as to materials to be used, control methods, standard operating procedures, and any other such material that is reasonably necessary or useful to enable Biogen or its designee to manufacture such Acquired Antibodies and Products;

(II) have the appropriate employees, representatives and consultants of the applicable Affiliate or Kiniksa Third Party manufacturer meet with employees of Biogen or its designee at the facilities of Biogen or its designee, from time to time as reasonably requested by Biogen, to assist with the working up and use of the process to manufacture such Acquired Antibodies and Products and with the training of Biogen's or its designee's personnel to the extent reasonably necessary or useful to enable Biogen or its designee to manufacture such Acquired Antibodies and Products; and

(III) take such steps as are reasonably necessary to assist Biogen or its designee in obtaining or varying any necessary Regulatory Approval with respect to Biogen's or its designee's manufacture of such Acquired Antibodies and Products; and

(IV) provide such other assistance as Biogen may reasonably request to enable Biogen or its designee to manufacture such Acquired Antibodies and Products;

(V) each Party shall take such actions as may be reasonably necessary to complete the successful transfer to Biogen of the Acquired Antibodies and Products; and

(VI) each Party shall promptly return all Confidential Information of the other Party that are not subject to a continuing license hereunder; provided, that, each Party may retain one copy of the Confidential Information of the other Party in its archives solely for the purpose of establishing the contents thereof and ensuring compliance with its obligations hereunder.

For purposes of clarity, and without limiting Biogen's obligations under Section 8.3(e)(iii), the costs and expenses incurred by Kiniksa in undertaking the actions set forth in subsection (i) through (vi) above shall be [***]; and

(f) on and after the effective date of such termination, all Acquired

Know-How and Acquired Patent Rights shall be considered Confidential Information of, and the property of, Biogen and the terms set forth in Section 7, and all related obligations of Biogen in Section 7, shall apply *mutatis mutandis* to Kiniksa.

8.4 **Kiniksa Rights in Lieu of Termination**. If Kiniksa has the right to terminate this Agreement pursuant to Section 8.2 for Biogen's material breach, Kiniksa may elect to either

(i) terminate this Agreement (in which case the provisions of Section 8.3 shall apply) or (ii) continue this Agreement, subject to the following provisions which shall be effective upon Kiniksa's notice of such election pursuant to this clause (ii) (the "***Kiniksa Election Notice***"):

(a) the licenses and rights granted by Biogen to Kiniksa pursuant to Section 2.5.1 shall remain in effect;

(b) milestone and royalty payments due to Biogen pursuant to Sections5.2 and 5.3 on and after the effective date of termination shall be [***].

(c) Nothing herein shall limit Kiniksa's rights to pursue damages pursuant to a claim under this Agreement. Except to the extent provided in this Section 8.4, this Agreement shall remain in full force and effect.

8.5 **Surviving Obligations**. The following portions of this Agreement shall survive termination or expiration of this Agreement: Sections 2.3 (with respect to Assumed Liabilities prior to the effective date of termination), 2.4, 5.3.5, 5.5, 5.6, 5.7, 8.3, 8.4, 8.5, 10.1 (as applicable) and 10.3, 10.4, 10.5, 10.6, 10.7, 10.10, and Articles 1 (as applicable), 7 and 9 (for the time periods set forth therein). All other portions of and obligations under this Agreement shall terminate (including Section 3.4) upon expiration or termination of this Agreement, except that expiration or termination of this Agreement shall not relieve the Parties of any liability or obligation which accrued hereunder prior to the effective date of such expiration or termination.

9. SURVIVAL; INDEMNIFICATION; INSURANCE; LIMITATIONS

9.1 **Survival of Representations and Warranties**. Except for those representations and warranties contained in Sections 6.2.1, 6.2.4(h) and 6.2.4(i) of this Agreement (which shall survive indefinitely), (a) those representations and warranties contained in Sections 6.2.2 and 6.2.3 of this Agreement shall continue in full force and effect for a period of twelve (12) months from the Effective Date and (b) those representations and warranties contained in Sections 6.2.4 of this Agreement shall continue in full force and effect for a period of eighteen (18) months from the Effective Date. Any claims for indemnification under Section 9.2 or Section 9.3 asserted in writing as provided for in this Article 9 prior to such expiration date, if any, applicable to the representation or warranty with respect to which such claim for indemnification is made shall survive until finally resolved and satisfied in full. No Third Party other than the Indemnified Parties shall be a Third Party or other beneficiary of any representations, warranties, covenants and agreements in this Agreement and no such Third Party shall have any

rights of contribution with respect to such representations, warranties, covenants or agreements or any matter subject to or resulting in indemnification under this Article 9 or otherwise. The representations, warranties, covenants and agreements set forth in this Agreement and in the Patent Assignment Agreement shall not be affected or diminished in any way by any investigation (or failure to investigate) at any time by or on behalf of the Party for whose benefit such representations, warranties, covenants and agreements were made.

9.2 **Indemnification by Kiniksa**. Subject to Sections 9.4 and 9.7, Kiniksa agrees to defend Biogen, its Affiliates and its (and its Affiliates') directors, officers, employees and agents (the "***Biogen Indemnified Parties***") at Kiniksa's cost and expense, and will indemnify and hold Biogen and the other Biogen Indemnified Parties harmless from and against any claims, losses, costs, damages, fees or expenses (including legal fees and expenses) (collectively, "***Losses***") resulting from any claims (including Third Party and product liability claims), actions or demands (collectively "***Claims***") arising out of or otherwise relating to:

(a) the negligence or willful misconduct of Kiniksa in connection with Kiniksa's performance of this Agreement;

(b) the material breach by Kiniksa of this Agreement, any Assigned Contract or the applicable terms of any Retained Contract, including any of the representations or warranties made hereunder by Kiniksa;

(c) the Exploitation of any Acquired Antibody or Product by or on behalf of Kiniksa or its Affiliates on or after the Effective Date; or

(d) the use by or on behalf of Kiniksa in any Clinical Trial of any materials provided by Biogen pursuant to Section 2.6.1.

except, in each case, to the extent such Losses arise out of or relate to such subsection (a), (b), (c) or (d) of Section 9.3. In the event of any such Claim against the Biogen Indemnified Parties by a Third Party, Biogen shall promptly notify Kiniksa in writing of the Claim (provided, that, any failure or delay to so notify Kiniksa shall not excuse any obligations of Kiniksa except to the extent Kiniksa is actually prejudiced thereby) and Kiniksa shall solely manage and control, at its sole expense, the defense of the Claim and its settlement; provided, that, Kiniksa shall not settle any such Claim without the prior written consent of Biogen if such settlement does not include a complete release of Biogen Indemnified Parties from liability or if such settlement would involve undertaking an obligation (including the payment of money by a Biogen Indemnified Party), would bind or impair a Biogen Indemnified Party, or includes any admission of wrongdoing or that any intellectual property or proprietary right of Biogen is invalid or unenforceable. The Biogen Indemnified Parties shall cooperate with Kiniksa and may, at their option and expense, be represented in any such action or proceeding by counsel of their own choosing. With respect to any Claim subject to indemnification under this Section 9.2: (i) both Kiniksa and the Biogen Indemnified Parties, as the case may be, shall keep the other Person fully informed of the status of such Claim and any related proceedings at all stages thereof where

such Person is not represented by its own counsel, (ii) the Parties agree (each at its own expense) to render to each other such assistance as they may reasonably require of each other and to cooperate in good faith with each other in order to ensure the proper and adequate defense of any such Claim and (iii) the Parties agree to cooperate in such a manner as to preserve in full (to the extent possible) the confidentiality of all Confidential Information and information protected by the attorney-client and work-product privileges in any such action or proceeding.

9.3 **Indemnification By Biogen**. Subject to Sections 9.4 and 9.7, Biogen agrees to defend Kiniksa, its Affiliates and its (and its Affiliates') directors, officers, employees and agents (the "***Kiniksa Indemnified Parties***") at Biogen's cost and expense, and will indemnify and hold Kiniksa and the other Kiniksa Indemnified Parties harmless from and against any Losses resulting from any Claims arising out of or otherwise relating to:

(a) the negligence or willful misconduct of Biogen or its Affiliates in connection with such parties' performance of this Agreement;

(b) the material breach by Biogen of this Agreement including any of the representations or warranties made hereunder by Biogen;

(c) the Exploitation of any Acquired Antibody or Product by or on behalf of Biogen or its Affiliates prior to the Effective Date; or

(d) the Exploitation of any Acquired Antibody or Product by or on behalf of Biogen or its Affiliates following termination of this Agreement and reversion of rights pursuant to Section 8.3.

except, in each case, to the extent such Losses arise out of or relate to subsections (a), (b), or (c) of Section 9.2. In the event of any such Claim against the Kiniksa Indemnified Parties by a Third Party, Kiniksa shall promptly notify Biogen in writing of the Claim (provided, that, any failure or delay to so notify Biogen shall not excuse any obligation of Biogen except to the extent Biogen is actually prejudiced thereby) and Biogen shall solely manage and control, at its sole expense, the defense of the Claim and its settlement; provided, that, Biogen shall not settle any such Claim without the prior written consent of Kiniksa if such settlement does not include a complete release of the Kiniksa Indemnified Parties from liability or if such settlement would involve undertaking an obligation (including the payment of money by an Kiniksa Indemnified Party), would bind or impair an Kiniksa Indemnified Party, or includes any admission of wrongdoing or that any intellectual property or proprietary right of Kiniksa is invalid or unenforceable. The Kiniksa Indemnified Parties shall cooperate with Biogen and may, at their option and expense, be represented in any such action or proceeding by counsel of their own choosing. With respect to any Claim subject to indemnification under this Section 9.3: (i) both Biogen and the Kiniksa Indemnified Parties, as the case may be, shall keep the other Person fully informed of the status of such Claim and any related proceedings at all stages thereof where such Person is not represented by its own counsel, (ii) the Parties agree (each at its own expense) to render to each other such assistance as they may reasonably require of each other and to

cooperate in good faith with each other in order to ensure the proper and adequate defense of any such Claim and (iii) the Parties agree to cooperate in such a manner as to preserve in full (to the extent possible) the confidentiality of all Confidential Information and information protected by the attorney-client and work-product privileges in any such action or prosecution.

9.4 **Limitation on Indemnification**. Notwithstanding anything to the contrary in Section 9.2 or 9.3, (a) no Indemnifying Party shall have any liability to the corresponding Indemnified Parties under Section 9.2 or Section 9.3 until the aggregate Losses of such Indemnified Parties exceed $50,000, after which the Indemnified Parties shall be entitled to all such Losses; and (b) Kiniksa's recourse against Biogen with respect to any right to indemnification under Sections 9.3 (a), (b) or (c) shall be limited in amount to the lesser of (i) the aggregate amount of all payments made by Kiniksa to Biogen hereunder determined at the time of payment of any such indemnification Claim and (ii) $41,000,000, except in the cases of (A) fraud, willful misconduct or intentional misrepresentation or (B) with respect to any Excluded Liability, in which case there shall be no limit.

9.5 **Sole Remedy**. Except to the extent that a claim involves fraudulent or willful misconduct or intentional misrepresentation, the sole and exclusive remedy for any material breach or alleged material breach of any representation, warranty or covenant shall be indemnification in accordance with this Article 9.

9.6 **Insurance**. Kiniksa will maintain, at its cost, reasonable insurance against liability and other risks associated with its activities contemplated by this Agreement, provided, that, if Kiniksa is engaged in the Development of Acquired Antibodies or Products hereunder, Kiniksa will maintain, in force from thirty (30) days prior to the Initiation of any Clinical Trial, a clinical trials/product liability insurance policy providing coverage of at least [***] per claim and [***] annually in the aggregate; provided, further, that, if Kiniksa Commercializes any Product, such coverage shall be increased to at least [***] at least thirty (30) days prior to the date of anticipated First Commercial Sale of such Product. Kiniksa shall provide thirty (30) days advance written notice to Biogen of the termination, cancellation or material alteration of the terms or conditions of its insurance policies. If such policies are written on a claims made basis, they shall remain in effect for a minimum period of five (5) years after the termination or expiration of this Agreement and shall not be cancelled or subject to a reduction of coverage without the prior written authorization of Biogen. Upon Biogen's written request, Kiniksa shall provide Biogen certified copies of Kiniksa's insurance policies to evidence the purchase and/or maintenance of such policies. Maintenance of such insurance coverage shall not relieve Kiniksa of any responsibility under this Agreement for damages in excess of insurance limits or otherwise.

9.7 **LIMITATION OF DAMAGES**. IN NO EVENT SHALL EITHER PARTY BE LIABLE HEREUNDER TO THE OTHER PARTY FOR ANY PUNITIVE, RELIANCE, INDIRECT, SPECIAL, INCIDENTAL OR CONSEQUENTIAL DAMAGES (INCLUDING LOST REVENUE, LOST PROFITS, OR LOST SAVINGS) HOWEVER CAUSED AND UNDER ANY THEORY, EVEN IF IT HAS NOTICE OF THE POSSIBILITY OF SUCH DAMAGES. THE LIMITATIONS SET FORTH IN THIS SECTION 9.7 SHALL NOT

APPLY WITH RESPECT TO (A) ANY BREACH OF ARTICLE 7 OR (B) THE INTENTIONAL MISCONDUCT OR FRAUD OF A PARTY. NOTHING IN THIS SECTION 9.7 IS INTENDED TO LIMIT OR RESTRICT THE INDEMNIFICATION RIGHTS OR OBLIGATIONS OF A PARTY UNDER THIS ARTICLE 9 WITH RESPECT TO ANY DAMAGES PAID BY THE OTHER PARTY TO A THIRD PARTY IN CONNECTION WITH A THIRD PARTY CLAIM.

10. MISCELLANEOUS

10.1 **Entire Agreement; Amendment**. This Agreement, all Schedules and Exhibits attached to this Agreement, and the Patent Assignment Agreement constitute the entire agreement between the Parties as to the subject matter hereof. Except as set forth in this Section 10.1, (i) all prior and contemporaneous negotiations, representations, warranties, agreements, statements, promises and understandings with respect to the subject matter of this Agreement are hereby superseded and merged into, extinguished by and completely expressed by this Agreement and the Patent Assignment Agreement and (ii) none of the Parties shall be bound by or charged with any written or oral agreements, representations, warranties, statements, promises or understandings not specifically set forth in this Agreement. No amendment, supplement or other modification to any provision of this Agreement shall be binding unless in writing and signed by both Parties. Notwithstanding the foregoing, except with respect to any rights and obligations of the Parties with respect to the Acquired Know-How or the Acquired Patent Rights which shall be governed solely by this Agreement, (i) all rights and obligations of the Parties that arose under the CDA during the period commencing on the CDA Effective Date and continuing through the Effective Date, including any dispute or alleged breach by a Party of any of the terms of the CDA during such period, shall be governed solely by the terms of the CDA, (ii) the terms and conditions of the CDA shall survive solely for the limited purposes set forth in subsection (i) above and (iii) the CDA shall otherwise terminate as of the Effective Date.

10.2 **Section 365(n) of the Bankruptcy Code**. All rights and licenses granted under or pursuant to any section of this Agreement are, and shall otherwise be deemed to be, for purposes of Section 365(n) of the U.S. Bankruptcy Code, licenses of rights to "intellectual property" as defined under Section 101(35A) of the U.S. Bankruptcy Code to the extent permitted thereunder. The Parties shall retain and may fully exercise all of their respective rights and elections under the U.S. Bankruptcy Code. Upon the bankruptcy of Biogen, Kiniksa shall further be entitled to a complete duplicate of (or complete access to, as appropriate) any such intellectual property, and such, if not already in its possession, shall be promptly delivered to Kiniksa, unless Biogen elects to continue, and continues, to perform all of its obligations under this Agreement.

10.3 **Governing Law; Jurisdiction**. This Agreement and its effect are subject to and shall be construed and enforced in accordance with the law of the State of New York, without regard to its conflicts of laws that would require the application of any other Law. Each of the Parties hereby irrevocably and unconditionally consents to submit to the exclusive jurisdiction of the federal courts located in the Eastern District of the State of New York for any matter

arising out of or relating to this Agreement and the transactions contemplated hereby, and agrees not to commence any litigation relating thereto except in such courts. Each of the Parties hereby irrevocably and unconditionally waives any objection to the laying of venue of any matter arising out of this Agreement or the transactions contemplated hereby in the courts of the State of New York and hereby further irrevocably and unconditionally waives and agrees not to plead or claim in any such court that any such matter brought in any such court has been brought in an inconvenient forum. The Parties agree that a final judgment in any such matter shall be conclusive and may be enforced in other jurisdictions by suits on the judgment or in any other manner provided by law. Any proceeding brought by either Party under this Agreement shall be exclusively conducted in the English language.

10.4 **Notice**. All notices or communication required or permitted to be given by either Party hereunder shall be deemed sufficiently given if mailed by registered mail or certified mail, return receipt requested, or sent by overnight courier, such as Federal Express, to the other Party at its respective address set forth below (including a copy as designated below) or to such other address as one Party shall give notice of to the other from time to time hereunder. Mailed notices shall be deemed to be received on the third (3rd) Business Day following the date of mailing. Notices sent by overnight courier shall be deemed received the following Business Day.

If to Kiniksa: Kiniksa Pharmaceuticals, Ltd.

Clarendon
House 2
Church Street

Hamilton HM
11 Bermuda

Attention: Chief Legal Officer

With copies to: Kiniksa Pharmaceuticals Corp.

15 Walnut Street

Wellesley, MA 02481
Attention: Chief Legal
Officer

and: Latham & Watkins
LLP John Hancock
Tower 27th Floor

200 Clarendon Street

Boston, MA 02116

Attn: Johan Brigham

If to Biogen: Biogen MA
Inc. 225
Binney Street

Cambridge, MA 02142
Attn: General Counsel

10.5 **Compliance With Law; Severability**. Nothing in this Agreement shall be construed to require the commission of any act contrary to Law. If any one or more provisions of this Agreement is held to be invalid, illegal or unenforceable, the affected provisions of this Agreement shall be curtailed and limited only to the extent necessary to bring it within the applicable legal requirements and the validity, legality and enforceability of the remaining provisions of this Agreement shall not in any way be affected or impaired thereby.

10.6 **Successors and Assigns**. This Agreement shall bind and inure to the benefit of the successors and permitted assigns of the Parties hereto. Neither this Agreement nor any right, interest or obligation of a Party hereunder (including, with respect to Kiniksa, any of the Acquired Patent Rights or Acquired Know-How) may be assigned by either Party without the written consent of the other Party, except that each Party may assign this Agreement and the rights, obligations and interests of such Party under this Agreement (a) in whole or in part, to any of its Affiliates, or (b) subject to Kiniksa's and its Affiliates' obligations with respect to ROFN Transactions pursuant to Section 3.4, in whole, but not in part, to any purchaser of all or substantially all of its assets or all or substantially all of its assets to which this Agreement relates or to the purchaser of shares representing a majority of its common stock voting rights or to the surviving corporation resulting from any merger, consolidation, share exchange or other similar transaction; provided, that, (i) the assigning Party will provide the other Party with prompt written notice of assignment, (ii) the permitted assignee will assume all obligations of its assignor under this Agreement and the Patent Assignment Agreement (or as related to the assigned part where a partial assignment to an Affiliate), (iii) unless expressly so agreed in writing by the Parties, no permitted assignment will relieve the assignor of liability under this Agreement or the Patent Assignment Agreement, and (iv) any attempted assignment in contravention of this Section 10.6 shall be void.

10.7 **Waivers**. A Party's consent to or waiver, express or implied, of any other Party's breach of its obligations hereunder shall not be deemed to be or construed as a consent to or waiver of any other breach of the same or any other obligations of such breaching Party. A Party's failure to complain of any act, or failure to act, by the other Party, to declare the other Party in default, to insist upon the strict performance of any obligation or condition of this Agreement or to exercise any right or remedy consequent upon a breach thereof, no matter how long such failure continues, shall not constitute a waiver by such Party of its rights hereunder,

of any such breach, or of any other obligation or condition. A Party's consent in any one instance shall not limit or waive the necessity to obtain such Party's consent in any future instance and in any event no consent or waiver shall be effective for any purpose hereunder unless such consent or waiver is in writing and signed by the Party granting such consent or waiver.

10.8 **Force Majeure**. Except for the obligation to pay money when due, neither Party shall be liable to the other for failure or delay in the performance of any of its obligations under this Agreement for the time and to the extent such failure or delay is caused by a Force Majeure. The Party affected by the Force Majeure shall provide the other Party in writing with full particulars thereof as soon as it becomes aware of the same (including its best estimate of the likely extent and duration of the interference with its activities), and will use commercially reasonable efforts to overcome the difficulties created thereby and to resume performance of its obligations as soon as practicable. For the avoidance of doubt, under no circumstances shall the alleged or actual inability to pay money be considered an event of Force Majeure.

10.9 **No Third Party Beneficiaries**. Nothing in this Agreement shall be construed as giving any Person, other than the Parties hereto and their successors and permitted assigns, any right, remedy or claim under or in respect of this Agreement or any provision hereof, except for the provisions of Article 9 (with respect to which the Persons to which Article 9 applies shall be Third Party beneficiaries in accordance with Article 9).

10.10 **Headings; Schedules and Exhibits**. Article and Section headings used herein are for convenient reference only, and are not a part of this Agreement. All Schedules and Exhibits are incorporated herein by this reference.

10.11 **Counterparts**. This Agreement may be executed in counterparts by a single Party, each of which when taken together shall constitute one and the same agreement, and may be executed through the use of facsimiles or .pdf documents.

10.12 **Further Assurances**. From time to time after the Effective Date, and for no further consideration (except as expressly set forth in Section 2.5 and Section 2.6), Biogen shall execute, acknowledge and deliver such assignments, transfers, consents, assumptions and other documents and instruments and take such other actions as may be necessary or desirable to consummate and make effective the transactions contemplated by this Agreement.

(Signature Page Immediately to Follow)

IN WITNESS WHEREOF, this Agreement has been executed by the Parties hereto all as of the date first above written.

KINIKSA PHARMACEUTICALS, LTD.

By: /s/ Thomas Beetham
Name: Thomas Beetham
Title: Executive Vice President

BIOGEN MA INC.

By: /s/ John McDonald
Name: John McDonald
Title: VP, Business Development

[Signature Page to Asset Purchase Agreement]

Schedule A

Purchased Assets

1. **Acquired Patent Rights**

[***]

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2. Acquired Know-How

[***]

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3. **Assigned Contracts**

[***]

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4. Inventory

A. [***]

[REDACTED]

B. [***]

C. [***]

Schedule B

Selected Obligations Under Sigma Agreement

[***]

Schedule C

Determination of Applicable Multiplier

[***]

Schedule D

Background Licensed Patent Rights

Publication Number	Title	Application Date	Publication Date	Inventor(s)	All IPC

[***]

Schedule E

Retained Contracts

1. Non-Exclusive Biological Material License Agreement by and between American Type Culture Collection (ATCC) and Biogen MA Inc., effective August 12, 2015 (the "**ATCC Agreement**").

2. Non-Exclusive License Agreement by and between SIGMA-ALDRICH CO. LLC (Sigma) and Biogen Idec MA, Inc., effective September 24, 2014 (the "**Sigma Agreement**").

Retained Contracts Payments

[***]

Schedule F

Description of BIIB069

[***]

Schedule G

Completion of Technology Transfer

[***]

Schedule H

Individuals for Purposes of Knowledge

[***]

Schedule I

Exceptions to Biogen Obligations/Representations in Sections 2.6.6 and 6.2.4(g) and (l)

[***]

Exhibit A

Form of Patent Assignment Agreement

PATENT ASSIGNMENT AGREEMENT

THIS PATENT ASSIGNMENT AGREEMENT (this "Assignment") is being entered into by and between Biogen MA Inc., a Massachusetts corporation ("Assignor"), and Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company ("Assignee").

WHEREAS, Assignor is an owner of the patent rights listed on Attachment A (collectively, the "Patents"); and

WHEREAS, Assignor has agreed with Assignee for the transfer to Assignee of all of Assignor's right, title and interest in and to said Patents and the inventions therein pursuant to a certain Asset Purchase Agreement between Assignor and Assignee, dated as of September , 2016 (the "Asset Purchase Agreement").

NOW THEREFORE, pursuant to such Asset Purchase Agreement and in consideration of the mutual covenants, agreements, representations and warranties contained herein and in the Asset Purchase Agreement, Assignor hereby sells, assigns, transfers, and sets over to Assignee or its heirs, successors, assigns, or other legal representatives the full and entire right, title, and interest in and to the Patents and the inventions therein, including the right of Assignee or its heirs, successors, assigns, or other legal representatives to: (a) file any nonprovisional patent applications and to otherwise seek any patent in the United States and any foreign jurisdiction claiming priority to the provisional patent application; (b) file any and all divisional, continuation, and continuation-in-part applications claiming priority to the nonprovisional patent application; and (c) seek reissues, reexaminations, adjustments, or extensions of any patent claiming priority to the provisional application which the Assignee may hold and enjoy as fully and entirely as Assignor would have had this assignment and sale not been made and Assignor acknowledges that Assignee has all rights under all applicable intellectual property treaties and conventions and the full benefits thereof and all rights, privileges and advantages appertaining thereto, TO HOLD the same unto and to the use of Assignee, its successors and assigns during the residue of the respective terms for which the said Patents were or will be granted and during any such terms, and for any and all rights extending from, including any divisions, continuations, continuations-in-part, reissues, reexaminations adjustments and extensions;

AND, for the same consideration, Assignor hereby covenants and agrees to and with Assignee, its successors, legal representatives and assigns that Assignor will sign all papers and documents, take all lawful oaths, and do all acts necessary or required to be done for the

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recordation of this assignment of the Patents and the inventions therein and other such acts that may be necessary or desirable to perfect the title to, including the invention, the provisional patent application, any application or applications claiming priority thereto and any patent or patents that may be obtained therefrom and Assignor further agrees to ratify and hereby ratifies any acts of Assignee in applying for a patent in Assignee's own name in any jurisdiction where such procedure is proper and agrees to execute or have executed any documents or assignments where it is necessary that they be executed by the inventor(s) and Assignor further agrees to execute assignments of any patent applications claiming the invention, any patent applications filed which claim priority to the provisional patent application, and any patents issuing from such patent applications to Assignee;

AND, Assignor represents and warrants that Assignor has the full right to convey the entire interest of the Patents, the inventions therein and the applications and has not granted any rights inconsistent with the rights granted in this Assignment.

Executed as of this 7th day of September, 2016.

ASSIGNOR:

BIOGEN MA INC.

By: _____
Name:
Title:

ASSIGNEE:

KINIKSA PHARMACEUTICALS, LTD.

By: _____
Name:
Title:

ATTACHMENT A

Assigned Patents

[***]

AMENDMENT NO. 1 TO
ASSET PURCHASE AGREEMENT

This Amendment No. 1 to Asset Purchase Agreement (this "***Amendment***") is dated as of July 31, 2017 (the "***Amendment Effective Date***") by and between Biogen MA Inc., a Massachusetts corporation ("***Biogen***"), and Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company ("***Kiniksa***"). Kiniksa and Biogen are sometimes referred to herein individually as a "***Party***" and collectively as the "***Parties***"). Terms used in this Amendment and not otherwise defined shall have the respective meanings set forth in the APA (as defined below).

WHEREAS, pursuant to the terms of the Asset Purchase Agreement (the "***APA***") dated as of September 7, 2016 (the "***APA Effective Date***"), by and between Biogen and Kiniksa, Biogen agreed to sell to Kiniksa, and Kiniksa agreed to purchase from Biogen, certain assets of Biogen used in or relating to BIIB069, all upon the terms and conditions set forth herein; and

WHEREAS, the Parties now wish to amend the APA to (a) provide for the transfer by Biogen to Kiniksa of certain quantities of an additional antibody of Biogen designated as BIIB22G11 and (b) clarify certain definitions under the APA related to BIIB22G11; and

WHEREAS, pursuant to Section 10.1 of the APA, no amendment, supplement or other modification to any provision of the APA shall be binding unless in writing and signed by both Parties.

NOW, THEREFORE, in consideration of the mutual covenants contained herein, and for other good and valuable consideration, the receipt and adequacy of which are hereby acknowledged, the Parties hereto, intending to be legally bound, hereby agree as follows:

Amendments to APA.

The following new definition is hereby inserted in alphabetical order in Section 1.1 of the APA:

"***BIIB22G11***" means the Antibody described on Schedule F-1 attached hereto."

The definition of "***Acquired Antibody***" in Section 1.1 of the APA is hereby deleted in its entirety and the following is hereby inserted in lieu thereof:

"***Acquired Antibody***" means (a) BIIB069, BIIB22G11 or any [***] Antibody that is Covered by one or more claims within the Acquired Patent Rights; (b) [***].

The definition of "***Inventory***" in Section 1.1 of the APA is hereby deleted in its entirety and the following is hereby inserted in lieu thereof:

"***Inventory***" means the inventory of BIIB069 and BIIB22G11 listed as Inventory in Part 4 of Schedule A attached hereto."

Section 3.2 of the APA is hereby deleted in its entirety and the following is hereby inserted in lieu thereof:

3.2 Diligence. Kiniksa shall use Commercially Reasonable Efforts to Develop and Commercialize Acquired Antibodies and Products [***] and to commit such resources (including employees, consultants, contractors, facilities, equipment and materials) as are necessary to conduct such Development and Commercialization activities. Kiniksa shall perform its obligations under this Agreement in good scientific manner and in compliance with all applicable Law. For purposes of clarity, with respect to each Development and/or Commercialization activity that will or would reasonably be expected to be submitted to a Regulatory Authority in support of a Regulatory Filing or Marketing Authorization, Kiniksa shall comply in all material respects with GLPs, GMPs or Good Clinical Practices (or, if and as appropriate under the circumstances, International Council for Harmonisation ("**ICH**") guidance or other comparable regulation and guidance of any Regulatory Authority in any country or region in the Territory)."

Part 4 of Schedule A to the APA is hereby amended by adding thereto the additional inventory of BIIB22G11 listed on Schedule A-1 attached hereto.

A new Schedule F-1 entitled "Description of BIIB22G11" is hereby added to the APA in the form of Schedule F-1 attached hereto.

Miscellaneous. The Parties hereby confirm and agree that, except as amended hereby, the APA remains in full force and effect and continues to be a binding obligation of the Parties. This Amendment may be executed in counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

[Remainder of page intentionally left blank]

IN WITNESS WHEREOF, the Parties hereto have executed this Amendment as of the Amendment Effective Date.

KINIKSA PHARMACEUTICALS, LTD.

By: /s/ Thomas Beetham
Name: Thomas Beetham
Title: Executive Vice President

BIOGEN MA INC.

By: /s/ John McDonald
Name: John McDonald
Title: Vice President, Business Development

Schedule A-1

Part 4 - **Inventory of BIIB22G11**

[***]

Schedule F-1

Description of BIIB22G11

[***]

Exhibit 10.8

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

LICENSE AGREEMENT

between

MEDIMMUNE, LIMITED

and

KINIKSA PHARMACEUTICALS, LTD.

Dated as of December 21, 2017

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

TABLE OF CONTENTS

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

Schedule 1.104 Transition Activites

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

Schedule 3.1 Purchased Assets

Schedule 8.2.3 Actions/Notices related to Exclusive Licensed Patents as at the Effective Date

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

LICENSE AGREEMENT

This License Agreement (the "**Agreement**") is made and entered into effective as of December 21, 2017 (the "**Effective Date**") by and between **MedImmune, Limited**, a limited liability company duly authorized and existing under the laws of England and Wales ("**MedImmune**") and **Kiniksa Pharmaceuticals, Ltd.,** a Bermuda exempted company ("**Licensee**"). MedImmune and Licensee are sometimes referred to herein individually as a "**Party**" and collectively as the "**Parties**."

RECITALS

WHEREAS, MedImmune owns and controls certain intellectual property rights with respect to the Licensed Compound (as defined herein) and Licensed Products (as defined herein) in the Territory (as defined herein); and

WHEREAS, MedImmune wishes to grant a license to Licensee, and Licensee wishes to take a license, under such intellectual property rights to develop and commercialize Licensed Products in the Territory, in each case in accordance with the terms and conditions set out below.

NOW, THEREFORE, in consideration of the premises and the mutual promises and conditions set out herein and other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, the Parties, intending to be legally bound, do hereby agree as follows:

ARTICLE 1
DEFINITIONS

Unless otherwise specifically provided herein, the following terms shall have the following meanings:

1.1. "**Affiliate**" means, with respect to a Party, any Person that, directly or indirectly, through one (1) or more intermediaries, controls, is controlled by or is under common control with such Party. For purposes of this definition, "control" and, with correlative meanings, the terms "controlled by" and "under common control with" means: (i) the possession, directly or indirectly, of the power to direct the management or policies of a business entity, whether through the ownership of voting securities, by contract relating to voting rights or corporate governance or otherwise; or (ii) the ownership, directly or indirectly, of more than fifty percent (50%) of the voting securities or other ownership interest of a business entity (or, with respect to a limited partnership or other similar entity, its general partner or controlling entity).

1.2. "**Agreement**" has the meaning set out in the preamble.

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

1.3. "**Anti-Corruption Laws**" means the U.S. Foreign Corrupt Practices Act, as amended, the UK Bribery Act 2010, as amended, and any other applicable anti-corruption laws and laws for the prevention of fraud, racketeering, money laundering or terrorism.

1.4. "**Applicable Law**" means applicable federal, state, regional, local or foreign laws, statues, codes or ordinance, rules and regulations, including any rules, regulations, guidelines or other requirements of the Regulatory Authorities, including all decisions of any Courts having the effect of law in each such jurisdiction, that may be in effect from time to time, including the FFDCA and the Anti-Corruption Laws.

1.5. "**Arbitrators**" has the meaning set out in Section 11.5.2.

1.6. "**Assumed Liabilities**" has the meaning set out in Section 3.3.

1.7. "**Auditor**" has the meaning set out in Section 5.10.

1.8. "**Bioequivalent**" means, a biological product that (i) is highly similar to the Licensed Product notwithstanding minor differences in clinically inactive components; and (ii) has no clinically meaningful differences between the biological product and the Licensed Product in terms of the safety, purity, and potency.

1.9. "**Biologics License Application**" or "**BLA**" means a Biologics License Application submitted to the FDA under subsection (a) or (k) of Section 351 of the FFDCA or any corresponding foreign application in the Territory, including, with respect to the European Union, a marketing authorization application filed with the EMA pursuant to the centralized approval procedure or with the applicable Regulatory Authority of a country in Europe (including Great Britain) with respect to the mutual recognition or any other national approval.

1.10. "**Biosimilar Product**" means with respect to a Licensed Product, a therapeutic product that (i) contains as an active ingredient any [***], (ii) is Bioequivalent to such Licensed Product, and (iii) is [***].

1.11. "**Board of Directors**" has the meaning set out in the definition of Change of Control.

1.12. "**Breaching Party**" has the meaning set out in Section 10.2.2.

1.13. "**Business Day**" means a day other than a Saturday or Sunday or a day on which commercial banking institutions in London, England or New York, NY USA are required to be closed.

Confidential Portions of this Exhibit marked as [***] have been omitted pursuant to a request for confidential treatment and have been filed separately with the Securities and Exchange Commission.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

1.14. "**Calendar Quarter**" means each successive period of three (3) calendar months commencing on January 1, April 1, July 1 and October 1, except that the first Calendar Quarter of the Term shall commence on the Effective Date and end on the day immediately prior to the first to occur of January 1, April 1, July 1 or October 1 after the Effective Date and the last Calendar Quarter shall end on the last day of the Term.

1.15. "**Calendar Year**" means each successive period of twelve (12) calendar months commencing on January 1 and ending on December 31, except that the first Calendar Year of the Term shall commence on the Effective Date and end on December 31 of the year in which the Effective Date occurs and the last Calendar Year of the Term shall commence on January 1 of the year in which the Term ends and end on the last day of the Term.

1.16. "**Change of Control**" with respect to a Party, shall be deemed to have occurred if any of the following occurs after the Effective Date:

(i) any "person" or "group" (as such terms are defined below) (i) is or becomes the "beneficial owner" (as defined below, except that a "person" or "group" shall be deemed to have "beneficial ownership" of all shares of capital stock or other equity interests if such person or group has the right to acquire, whether such right is exercisable immediately or only after the passage of time), directly or indirectly, of shares of capital stock or other interests (including partnership interests) of such Party then outstanding and normally entitled (without regard to the occurrence of any contingency) to vote in the election of the directors, managers or similar supervisory positions ("**Voting Stock**") of such Party representing fifty percent (50%) or more of the total voting power of all outstanding classes of Voting Stock of such Party or (ii) has the power, directly or indirectly, to elect a majority of the members of the Party's board of directors or similar governing body ("**Board of Directors**");

(ii) such Party enters into a merger, consolidation or similar transaction with another Person (whether or not such Party is the surviving entity) and as a result of such merger, consolidation or similar transaction (i) the members of the Board of Directors of such Party immediately prior to such transaction constitute less than a majority of the members of the Board of Directors of such Party or such surviving Person immediately following such transaction or (ii) the Persons that beneficially owned, directly or indirectly, the shares of Voting Stock of such Party immediately prior to such transaction cease to beneficially own, directly or indirectly, shares of Voting Stock of such Party representing at least a majority of the total voting power of all outstanding classes of Voting Stock of the surviving Person in substantially the same proportions as their ownership of Voting Stock of such Party immediately prior to such transaction;

(iii) such Party sells or transfers to any Third Party, in one or more related transactions, properties or assets representing all or substantially all of such Party's consolidated total assets to which this Agreement relates; or

(iv) the holders of capital stock of such Party approve a plan or proposal for the liquidation or dissolution of such Party.

For the purpose of this definition of Change of Control: (a) "**person**" and "**group**" have the meanings given such terms under Section 13(d) and 14(d) of the United States Securities Exchange Act of 1934 and the term "group" includes any group acting for the purpose of acquiring, holding or disposing of securities within the meaning of Rule 13d-5(b)(1) under the aforesaid Act; (b) a "**beneficial owner**" shall be determined in accordance with Rule 13d-3 under the aforesaid Act; and (c) the terms "**beneficially owned**" and "**beneficially own**" shall have meanings correlative to that of "beneficial owner."

1.17. "**Combination Product**" means a Licensed Product that is comprised of or contains the Licensed Compound as an active ingredient together with one (1) or more other active ingredients and is sold either as a fixed dose/unit or as separate doses/units in a single package.

1.18. "**Commercialization**" means any and all activities directed to the preparation for sale of, offering for sale of or sale of a Licensed Product, including activities related to Manufacturing for commercial sale, marketing, promoting, distributing and importing such Licensed Product and interacting with Regulatory Authorities regarding any of the foregoing. When used as a verb, "**to Commercialize**" and "**Commercializing**" means to engage in Commercialization and "**Commercialized**" has a corresponding meaning.

1.19. "**Commercialization Plan**" has the meaning set out in Section 4.3.2.

1.20. "**Commercially Reasonable Efforts**" means, with respect to the performance of Development, Commercialization and Manufacturing activities with respect to the Licensed Compound or a Licensed Product by Licensee, the carrying out of such activities using efforts and resources comparable to [***]. For purposes of the above, all relevant factors as measured by the facts and circumstances at the time such efforts are due shall be taken into account, including, as applicable and without limitation, mechanism of action; efficacy and safety; product profile; actual or anticipated Regulatory Authority approved labeling; the nature and extent of market exclusivity (including patent coverage, proprietary position and regulatory exclusivity); costs; time required for and likelihood of obtaining Regulatory Approval; expected competitive position of any such Licensed Product vis-à-vis other therapies that have been or are reasonably expected to be developed, marketed and sold or used for the same or similar indications; the presence of third-party Intellectual Property that is reasonably expected to impact the marketability of any such products; regulatory landscape; anticipated pricing and reimbursement for any such Licensed Product; and actual or projected profitability.

1.21. "**Confidential Information**" has the meaning set out in Section 7.1.

1.22. "**Control**" means, with respect to any item of Information, Regulatory Documentation, material, Patent or other Intellectual Property, possession of the right, whether directly or indirectly and whether by ownership, license or otherwise (other than by operation of the license and other grants in Section 3), to grant a license, sublicense or other right (including the right to reference Regulatory Documentation) to or under such Information, Regulatory Documentation, material, Patent or other Intellectual Property as provided for herein without violating the terms of any agreement with any Third Party.

1.23. "**Controlling Party**" has the meaning set out in Section 6.5.

1.24. "**Court**" means any court or arbitration tribunal of the United States, any domestic state, or any foreign country, and any political subdivision thereof.

1.25. **"Cover"** means, when referring to a Licensed Compound or Licensed Product with respect to a Patent, that, in the absence of a license granted to a Person under a claim included in such Patent, the practice by such Person of a specified activity with respect to such Licensed Compound or Licensed Product would infringe such claim.

1.26. "[***]" means the [***].

1.27. "**Development**" means all activities related to research, pre-clinical and other non-clinical testing, test method development and stability testing, toxicology, formulation, process development, manufacturing scale-up, qualification and validation, quality assurance/quality control, clinical studies, including Manufacturing in support thereof, statistical analysis and report writing, the preparation and submission of Biologics License Applications, regulatory affairs with respect to the foregoing and all other activities necessary or reasonably useful or otherwise requested or required by a Regulatory Authority as a condition or in support of obtaining or maintaining a Regulatory Approval. When used as a verb, "**Develop**" means to engage in Development.

1.28. "**Development Plan**" shall mean the [***] rolling plan for the Development of the Licensed Product for Commercialization in the Field in the Territory, which plan shall include the overall strategies and estimated timelines for Developing and submitting Regulatory Approvals for such Product within such [***] period in the Field in the Territory (inclusive of planned pre-clinical research and clinical development directed to the Licensed Product).

1.29. "**Dispute**" has the meaning set out in Section 11.5.

1.30. "**Dollars**" or "**$**" means United States Dollars.

1.31. "**Effective Date**" has the meaning set out in the preamble hereto.

1.32. "**EMA**" means the European Medicines Agency and any successor agency thereto.

1.33. "**Enforcing Party**" has the meaning set out in Section 6.3.2.

1.34. "**European Union**" means the economic, scientific and political organization of member states as it is constituted as of the Effective Date.

1.35. "**Excluded Liabilities**" has the meaning set out in Section 3.4.

1.36. "**Excluded Technology**" means certain materials and know-how relating to MedImmune's [***], and certain related trade secrets.

1.37. "**Exclusive Licensed Technology**" means the Exclusive Licensed Patents and the Exclusive Licensed Know How.

1.38. "**Exclusive Licensed Know How**" means all Information Controlled by MedImmune or any of its Affiliates as of the Effective Date that solely and/or exclusively relates to, the Licensed Compound or a Licensed Product (including without limitation, any Information sublicensed under [***]). For clarity, Exclusive Licensed Know How excludes the Excluded Technology and Non-Exclusively Licensed Know How.

1.39. "**Exclusive Licensed Patents**" means the (i) Patents listed on Schedule 1.39, which include without limitation, the Patents in-licensed under [***], (ii) all patent applications filed either from and claiming priority to the foregoing patents, patent applications or provisional applications of clause (i) or filed from an application claiming priority to any of the patent applications in this clause (ii), including divisionals, continuations, continuations-in-part, provisionals, converted provisionals and continued prosecution applications; (iii) all patents that have issued or in the future issue from the foregoing patent applications; and (iv) any and all extensions or restorations by existing or future extension or restoration mechanisms, including revalidations, reissues, re-examinations and extensions (including any supplementary protection certificates and the like) of the foregoing patents or patent applications.

1.40. "**Exploit**" means to make, have made, import, use, sell or offer for sale, including to research, Develop, Commercialize, register, Manufacture, have Manufactured, hold or keep (whether for disposal or otherwise), have used, export, transport, distribute, promote, market or have sold or otherwise dispose of a compound or product. "**Exploitation**" means the act of Exploiting a compound, product or process.

1.41. "**FDA**" means the United States Food and Drug Administration and any successor agency thereto.

1.42. "**FFDCA**" means the United States Food, Drug, and Cosmetic Act, as amended from time to time, together with any rules, regulations and requirements promulgated thereunder (including all additions, supplements, extensions and modifications thereto).

1.43. "**Field**" means all human diagnostic, prophylactic and therapeutic uses.

1.44. "**First Commercial Sale**" means, with respect to a Licensed Product and a country, the first sale for monetary value for use or consumption by the end user of such Licensed Product in such country after Regulatory Approval for such Licensed Product has been obtained in such country; provided that the following shall not constitute a First Commercial Sale: (i) any sale to an Affiliate or licensee (unless the Affiliate or licensee is the last entity in the distribution chain of the Licensed Product), or (ii) any transfers of a Licensed Product without consideration or for nominal consideration for use in any clinical trial, or for any bona fide charitable, compassionate use or indigent patient program purpose where Licensed Products are sold at or below cost of goods sold or as a sample.

1.45. "**FTE**" means the equivalent of the work of one employee full time for one year consisting of at least a total of [***] weeks or [***] hours per year (excluding vacations and holidays). For purposes of clarity, no one person shall be permitted to account for more than one FTE.

1.46. "**FTE Rate**" means $[***] per FTE per year.

1.47. "**Governmental Entity**" means any court, tribunal, arbitrator, Regulatory Authority, agency, commission, department, ministry, official or other instrumentality of the United States or other country, or any supra-national organization, or any foreign or domestic, state, county, city or other political subdivision.

1.48. "**Government Official**" means (i) any Person employed by or acting on behalf of a Governmental Entity, (ii) any political party, party official or candidate, (iii) any Person who holds or performs the duties of an appointment, office or position created by custom or convention, or (iv) any Person who holds himself out to be the authorized intermediary of any of the foregoing.

1.49. "**IND**" means (i) an investigational New Drug Application (as defined in the FFDCA and the regulations promulgated thereunder) or any successor application or procedure required to be filed with the FDA for authorization to commence clinical studies; (ii) its equivalent in other countries or regulatory jurisdictions before beginning clinical testing of a therapeutic product in humans in such country or region; and (iii) all supplements and amendments that may be filed with respect to the foregoing.

1.50. "**Indemnification Claim Notice**" has the meaning set out in Section 10.3.1.

1.51. "**Indemnified Party**" has the meaning set out in Section 9.3.1.

1.52. "**Indication**" means [***] Regulatory Approval (e.g., [***] such as (i) [***]; or (ii) [***], is not deemed to be a new indication for the purposes of this Agreement.

1.53. "**Information**" means all technical, scientific and other know-how and inventions, discoveries, improvements, information, trade secrets, business methods, knowledge, technology, means, methods, techniques, processes, practices, formulae, instructions, skills, techniques, procedures, experiences, ideas, technical assistance, designs, drawings, assembly procedures, computer programs, apparatuses, specifications, customer lists, cell lines, data, results and other material, including: biological, chemical, pharmacological, toxicological, pharmaceutical, physical and analytical, pre-clinical, clinical, safety, manufacturing and quality control data and tangible or intangible information, including study designs and protocols, assays and biological methodology, in each case (whether or not confidential, proprietary, patented or patentable) in written, electronic or any other form now known or hereafter developed.

1.54. "**Infringement**" has the meaning set out in Section 6.3.1.

1.55. "**Initiation**" means, with respect to a clinical study, the first dosing of the first human subject in such clinical study.

1.56. "**In-License Agreements**" means the licenses and other agreements entered into prior to the Effective Date by and between MedImmune or any of its Affiliates, on the one hand, and one (1) or more Third Parties on the other hand, that are necessary for, or were used in the Exploitation of the Licensed Compound and Licensed Product, and are listed on Schedule 1.56 (including without limitation, [***]).

1.57. **"Intellectual Property"** means any or all of the following and all rights in, arising out of, or associated therewith: (i) Patents; (ii) Information; (iii) all works of authorship, copyrights, copyrights registrations and applications therefor, and all other rights corresponding thereto throughout the world; (iv) all industrial designs and any registrations and applications therefor throughout the world; (v) all trade names, logos, common law trademarks and service marks, trademark and service mark registrations and applications therefor throughout the world and all goodwill associated therewith; (vi) all databases and data collections and all rights therein throughout the world; (vii) all moral and economic rights of authors and inventors, however denominated, throughout the world; (viii) all web addresses, sites and domain names and numbers; and (ix) any similar or equivalent rights to any of the foregoing anywhere in the world.

1.58. "**Inventory**" means the Licensed Compound and Licensed Product in its physical form as manufactured and currently stored by MedImmune, including without limitation, all non-cGMP drug substance, all cGMP drug substance, all finished drug

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product (including without limitation, placebo product), all process intermediates (including without limitation, the reference standards and cells lines), GMCSFRa surrogate antibodies (i.e., Licensed Compound that is not Mavriliumumab) and corresponding cell lines expressing such antibodies, analytical reagents, development cell banks, master and working cell banks, and cell lines to the extent related to the Licensed Compound or Licensed Product, the details and amounts of which is set out in Schedule 3.1.

1.59. "**Invoiced Sales**" has the meaning set out in the definition of "**Net Sales**."

1.60. "**Issuing Party**" has the meaning set out in Section 7.4.2.

1.61. "**Knowledge**" means the actual knowledge after performing a reasonably diligent investigation.

1.62. "**Liability**" means any and all debts, liabilities and obligations, whether known or unknown, asserted or unasserted, determinable or otherwise, accrued or fixed, absolute or contingent, liquidated or unliquidated, incurred or consequential, or matured or unmatured, including, without limitation, those arising under any Applicable Law, Litigation, order, or contract.

1.63. "**Licensee Intellectual Property**" means (i) all Intellectual Property, including any inventions, discoveries, Information, developments or modifications, whether or not patentable, that (a) are [***] related to the Licensed Compound or a Licensed Product for the Exploitation thereof [***], and (b) that are necessary to Develop, Manufacture, or Commercialize any Licensed Product, and (c) that are actually used during the Term by Licensee or its Affiliates in connection therewith, and (ii) all Regulatory Documentation (including any Regulatory Approvals) then owned or Controlled and/or created by Licensee or any of its Affiliates or its or their Sublicensees for the Exploitation of the Licensed Compound and Licensed Product(s) after the Effective Date.

1.64. "**Licensee Representatives**" has the meaning set out in Section 8.6.1.

1.65. "**Licensed Compound**" means (i) Mavrilimumab, a human monoclonal antibody targeting Granulocyte-Macrophage Colony Stimulating Factor Receptor Alpha (GMCSFRa) also known at MedImmune as CAM3001, and (ii) any other GMCSFRa antagonist Covered by one or more claims within the Licensed Patents.

1.66. **"Licensed Formulation Patents"** means any Patent claiming priority to U.S. patent application serial no. [***] related to the Licensed Compound [***]. For clarity, Licensed Formulation Patents are included in Non-Exclusive Licensed Patents, unless Licensee exercises the Option in Section 2.1(iii), in which case Licensed Formulation Patents are included in the Exclusive Licensed Patents.

1.67. "**Licensed Know How**" means the Exclusive Licensed Know How and the Non-Exclusive Licensed Know How.

1.68. "**Licensed Patents**" means the Exclusive Licensed Patents and the Non-Exclusive Licensed Patents.

1.69. "**Licensed Product**" means any product that is comprised of or contains the Licensed Compound.

1.70. "**Licensed Product Agreement**" means, with respect to a Licensed Product, any agreement entered into by and between Licensee or any of its Affiliates or its or their Sublicensees, on the one hand and one (1) or more Third Parties, on the other hand, that is necessary or reasonably useful for the Exploitation of such Licensed Product in the Field in the Territory, including (i) any agreement pursuant to which Licensee, its Affiliates or its or their Sublicensees receives any license or other rights to Exploit such Licensed Product, (ii) supply agreements pursuant to which Licensee, its Affiliates or its or their Sublicensees obtain or will obtain quantities of such Licensed Product, (iii) clinical trial agreements, (iv) contract research organization agreements, and (v) service agreements.

1.71 "**Licensed Technology**" means the Exclusive Licensed Technology and Non-Exclusive Licensed Technology.

1.71. "**Licensee**" has the meaning set out in the preamble hereto.

1.72. "**Licensee Representatives**" has the meaning set out in Section 8.6.1.

1.73. "**Litigation**" means any suit, action, arbitration, cause of action, claim, complaint, criminal prosecution, investigation, inquiry, demand letter, judicial, arbitration or other administrative proceeding, whether at law or at equity, before or by any Court, Governmental Entity, arbitrator or other tribunal.

1.74. "**Losses**" has the meaning set out in Section 9.1.

1.75. "**Manufacture**" and "**Manufacturing**" means all activities related to the production, manufacture, processing, filling, finishing, packaging, labeling, shipping and holding of a product or any intermediate thereof, including process development, process qualification and validation, scale-up, pre-clinical, clinical and commercial manufacture and analytic development, product characterization, stability testing, quality assurance and quality control.

1.76. "**Material Anti-Corruption Law Violation**" means a violation of an Anti-Corruption Law relating to the subject matter of this Agreement that would, if it were

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publicly known, in the reasonable view of MedImmune, have a material adverse effect on MedImmune or on the reputation of MedImmune because of its relationship with Licensee.

1.77. "**MedImmune**" has the meaning set out in the preamble hereto.

1.70 "**MedImmune Regulatory Documentation**" means Regulatory Documentation Controlled by MedImmune or any of its Affiliates as of the Effective Date relating to, but not exclusively relating to, the Licensed Compound or Licensed Product in the Field in the Territory.

1.71 "**Net Sales**" means, with respect to a Licensed Product for any period, the gross amounts invoiced by the Licensee or its Affiliates to Third Parties or Sublicensees for sales of the Licensed Product in the Territory (the "**Invoiced Sales**"), less the following deductions:

(i) trade, quantity, governmental or cash discounts, credits, adjustments or allowances, including those granted on account of price adjustments, billing errors, sales returns, rejected goods or damaged goods or goods otherwise not in saleable condition;

(ii) rebates and chargebacks allowed, given or accrued (including cash, Medicare, Medicaid, governmental and managed care rebates, hospital or other buying group chargebacks, and governmental taxes in the nature of a rebate based on usage levels or sales of the Licensed Product);

(iii) credits or allowances given or made for rejection, recall, return or wastage replacement of the Licensed Product;

(iv) taxes, duties or other governmental charges levied on or measured by the billing amount for Licensed Product (not offset or refunded, except in the case of value added taxes) assessed on the sale of the Licensed Product;

(v) any other similar and customary deductions that are consistent with U.S. GAAP; and

(vi) charges or allowances for transportation costs, customs, distribution expenses, special packaging and related insurance charges, freight and insurance charges, taken in accordance with Purchaser's standard practices, which charges or allowance will in no event exceed [***] of the amount arrived at after application of items (i) to (v) above.

For the avoidance of doubt, (a) in the case of any sale or other disposal of a Licensed Product between or among Licensee and its Affiliates for resale, invoiced sales and Net

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Sales shall be calculated only on the amount invoiced on the first arm's length sale thereafter to a Third Party; and (b) Net Sales shall not be imputed to transfers of Licensed Product (I) without consideration or for nominal consideration for use in any clinical trial or any other human studies reasonably necessary to comply with any Applicable Law or regulation or any request by a Regulatory Authority, (II) for any bona fide charitable, compassionate use or indigent patient or other similar program purpose where Licensed Products are sold at or below cost of goods sold, or (III) in commercially reasonable quantities as samples for promotional purposes.

In the event that a Licensed Product is sold in any country in the form of a Combination Product, Net Sales of such Combination Product shall be adjusted by multiplying actual Net Sales of such Combination Product in such country calculated pursuant to the foregoing definition of "Net Sales" by the fraction A/(A+B), where A is the average invoice price in such country of any Licensed Product that contains the same Licensed Compound(s) as such Combination Product as its sole active ingredient(s), if sold separately in such country and B is the average invoice price in such country of each product that contains active ingredient(s) other than the Licensed Compound(s) contained in such Combination Product as its sole active ingredient(s), if sold separately in such country; *provided* that the invoice price in a country for each Licensed Product that contains only the Licensed Compound(s) and each product that contains solely active ingredient(s) other than the Licensed Compound(s) included in the Combination Product shall be for a quantity comparable to that used in such Combination Product and of substantially the same class, purity and potency. If either such Licensed Product that contains the Licensed Compound(s) as its sole active ingredient or a product that contains an active ingredient (other than the Licensed Product) in the Combination Product as its sole active ingredient(s) is not sold separately in a particular country, the Parties shall negotiate in good faith a reasonable adjustment to Net Sales in such country that takes into account the medical contribution to the Combination Product of and all other factors reasonably relevant to the relative value of, the Licensed Compound(s), on the one hand and all of the other active ingredient(s), collectively, on the other hand.

Subject to the above, Net Sales shall be calculated in accordance with the standard internal policies and procedures of Licensee, its Affiliates or its or their Sublicensees, which must be in accordance with U.S. GAAP.

1.72. "**Non-Breaching Party**" has the meaning set out in Section 10.2.2.

1.72. "**Non-Exclusive In-License Agreement**" means In-License Agreements that are non-exclusively sublicensed to Licensee under Section 2.1(ii) and are listed on Schedule 1.56.

1.73. "**Non-Exclusive Licensed Know How**" means all Information Controlled by MedImmune or any of its Affiliates as of the Effective Date that is necessary for, or was

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used in, the Exploitation of the Licensed Compound or Licensed Products. For clarity, Non-Exclusive Licensed Know How excludes Excluded Technology.

1.74. "**Non-Exclusive Licensed Patents**" means all Patents Controlled by MedImmune as of the Effective Date that are necessary for, or were used in, the Exploitation of the Licensed Compound or Licensed Products, including without limitation, (i) Patents owned by MedImmune or its Affiliates set forth on Schedule 1.74, and (ii) any Patents sublicensed to Licensee under the Non-Exclusive In-License Agreements). For clarity, Non-Exclusive Licensed Patents excludes Exclusive Licensed Patents.

1.75. "**Non-Exclusive Licensed Technology**" means the Non-Exclusive Licensed Patents and Non-Exclusive Licensed Know How.

1.76. "**Option**" shall have the meaning set out in Section 2.1(iv).

1.77. "**Notice Period**" shall have the meaning set out in Section 10.2.2.

1.78. "**Party**" and "**Parties**" have the meaning set out in the preamble hereto.

1.79. "**Patents**" means: (i) all national, regional and international patents and patent applications, including provisional patent applications; (ii) all patent applications filed either from such patents, patent applications or provisional applications or from an application claiming priority to either of these, including divisionals, continuations, continuations-in-part, provisionals, converted provisionals and continued prosecution applications; (iii) any and all patents that have issued or in the future issue from the foregoing patent applications ((i) and (ii)), including utility models, petty patents, innovation patents and design patents and certificates of invention; (iv) any and all extensions or restorations by existing or future extension or restoration mechanisms, including revalidations, reissues, re-examinations and extensions (including any supplementary protection certificates and the like) of the foregoing patents or patent applications ((i), (ii) and (iii)); and (v) any similar rights, including so-called pipeline protection or any importation, revalidation, confirmation or introduction patent or registration patent or patent of additions to any of such foregoing patent applications and patents.

1.80. "**Payment**" has the meaning set out in Section 5.5.1.

1.81. "**Person**" means an individual, sole proprietorship, partnership, limited partnership, limited liability partnership, corporation, limited liability company, business trust, joint stock company, trust, unincorporated association, joint venture or other similar entity or organization, including a government or political subdivision, department or agency of a government.

1.82. "**Product Trademarks**" means the Trademark(s) used or to be used by Licensee or its Affiliates or its or their Sublicensees for the Commercialization of Licensed Products in the Territory (excluding, in any event, any corporate names and any Trademarks that consist of or include any corporate name or corporate logo of the Parties or their Affiliates or its or their (sub)licensees (or Sublicensees)).

1.70 "**Product Regulatory Documentation**" means Regulatory Documentation Controlled by MedImmune or any of its Affiliates as of the Effective Date relating [***] to the Licensed Compound or Licensed Product in the Field in the Territory.

1.83. "**Prosecuting Party**" has the meaning set out in Section 6.2.2.

1.84. "**Purchased Assets"** has the meaning set out in Section 3.1.

1.85. "**Release**" has the meaning set out in Section 7.4.2.

1.86. "**Regulatory Approval**" means, with respect to a country in the Territory, any and all approvals (including Biologics License Applications), licenses, registrations or authorizations of any Regulatory Authority necessary to commercially distribute, sell or market a Licensed Product in such country, including, where applicable, (i) pricing or reimbursement approval in such country, (ii) pre- and post-approval marketing authorizations (including any prerequisite Manufacturing approval or authorization related thereto) and (iii) labeling approval.

1.87. "**Regulatory Authority**" means any applicable supra-national, federal, national, regional, state, provincial or local regulatory agencies, departments, bureaus, commissions, councils or other government entities regulating or otherwise exercising authority with respect to the Exploitation of Licensed Compound or Licensed Products in the Territory, including the FDA in the United States and the EMA in the European Union.

1.88. "**Regulatory Documentation**" means: all (i) applications (including all INDs and Biologics License Applications), registrations, licenses, authorizations and approvals (including Regulatory Approvals); (ii) correspondence and reports submitted to or received from Regulatory Authorities (including minutes and official contact reports relating to any communications with any Regulatory Authority) and all supporting documents with respect thereto, including all adverse event files and complaint files; and (iii) clinical and other data contained or relied upon in any of the foregoing; in each case ((i), (ii) and (iii)) relating to the Licensed Compound or a Licensed Product.

1.89. "**Regulatory Exclusivity Period**" means, with respect to each Licensed Product in any country in the Territory, any period of data, market or other regulatory exclusivity (other than Patent exclusivity) granted or afforded by Applicable Law or by a Regulatory Authority in such country that confers exclusive marketing rights with respect to

such Licensed Product in such country and prevents another party from using or otherwise relying on any Regulatory Approval.

1.90. "**Retained Rights**" mean (i) with respect to the Licensed Compound and Licensed Products in the Field in the Territory, the rights of MedImmune, its Affiliates and its and their licensors, (sub)licensees and contractors to perform its and their obligations under this Agreement; (ii) with respect to the Excluded Technology the rights of MedImmune, its Affiliates and its and their licensors, (sub)licensees and contractors, to develop, obtain and maintain regulatory approvals for and to Exploit any compound or product, other than the Licensed Compound or Licensed Products, in any field (including the Field) anywhere in the Territory.

1.91. "**Reviewing Party**" has the meaning set out in Section 7.4.2.

1.92. "**Royalty Term**" means, with respect to each Licensed Product and each country in the Territory, the period beginning on the date of the First Commercial Sale of such Licensed Product in such country and ending on the latest to occur of: (i) the expiration date of the last to expire Valid Claim of a Patent included in the Exclusive Licensed Patents that Covers the Licensee's (or its Affiliate's or Subliensee's) manufacture, importation or sale of such Licensed Product in such country, (ii) the expiration of the Regulatory Exclusivity Period for such Licensed Product in such country, or (iii) the 10th anniversary of the First Commercial Sale of the Licensed Product in such country.

1.93. "**Senior Officer**" means, with respect to MedImmune, its Executive Vice President responsible for this Agreement, its Chief Executive Officer or President.

1.94. "**Sublicensee**" means a Person, other than an Affiliate, that is granted a sublicense by Licensee or its Affiliate under the grants in Section 2.1, as provided in Section 3.2.

1.95. **"Tax**" or "**Taxes**" means all income, excise, gross receipts, ad valorem, sales, use, employment, environmental, franchise, profits, gains, property, transfer, value added, payroll, escheat or abandoned property, intangibles or other taxes, fees, stamp taxes, duties, charges, levies or assessments of any kind whatsoever (whether payable directly or by withholding), together with any interest and any penalties, additions to tax or additional amounts imposed by any Governmental Entity with respect thereto, whether as a primary obligor, as a result of being a transferee, successor or a member of an affiliated, consolidated, unitary, combined or other group, by contract, pursuant to Applicable Law or otherwise.

1.96. "**Term**" has the meaning set out in Section 10.1.

1.97. "**Termination Notice**" has the meaning set out in Section 10.2.2.

1.98. "**Territory**" means worldwide.

1.99. "**Third Party**" means any Person other than MedImmune, Licensee and their respective Affiliates.

1.100. "**Third Party Claims**" has the meaning set out in Section 9.1.

1.101. "**Third Party Infringement Claim**" has the meaning set out in Section 6.4.

1.102. "**Third Party Patent Right**" has the meaning set out in Section 6.6.

1.103. "**Trademark**" means any word, name, symbol, color, shape, designation or any combination thereof, including any trademark, service mark, trade name, brand name, sub-brand name, trade dress, product configuration rights, program name, delivery form name, certification mark, collective mark, logo, tagline, slogan, design or business symbol, that functions as an identifier of source, origin or quality, whether or not registered, and all statutory and common law rights therein and all registrations and applications therefor, together with all goodwill associated with, or symbolized by, any of the foregoing.

1.104. "**Transition Activities**" means the activities to be conducted by MedImmune and applicable timeframes, to transfer the Licensed Technology and the Licensed Compound to Licensee as set out in Schedule 1.104.

1.105. **"Transition Services Agreement"** means a supplemental agreement to this Agreement that may be executed after the Effective Date, if required by the Parties, to more fully set out the Transition Activities.

1.106. "**United States**" or "**U.S.**" means the United States of America and its territories and possessions (including the District of Columbia and Puerto Rico).

1.107. "**U.S. GAAP**" means United States Generally Accepted Accounting Principles.

1.108. "**Valid Claim**" means (i) a claim of any issued and unexpired Patent whose validity, enforceability or patentability has not been affected by (a) irretrievable or unrevivable lapse, abandonment, revocation, dedication to the public or disclaimer or (b) a holding, finding or decision of invalidity, unenforceability or non-patentability by a court, governmental agency, national or regional patent office or other appropriate body that has competent jurisdiction, such holding, finding or decision being final and unappealable or unappealed within the time allowed for appeal or (ii) a claim of a pending Patent application that was filed and is being prosecuted in good faith and has not been abandoned or finally disallowed without the possibility of appeal or re-filing of the application; *provided, however*, that Valid Claim will exclude any such pending claim in any such Patent

application that has not been granted within [***] years after the earliest filing date from which such Patent application takes priority.

1.109. "**VAT**" has the meaning set out in Section 5.6.2.

1.110. "**Voting Stock**" has the meaning set out in the definition of "**Change of Control**."

ARTICLE 2
GRANT OF RIGHTS

2.1. Grants to Licensee. Medimmune hereby grants to Licensee:

(i) an exclusive (including with regard to MedImmune and its Affiliates) license (or sublicense), with the right to grant sublicenses through multiple tiers in accordance with Section 2.2, to the Exclusive Licensed Technology to Exploit the Licensed Compound and Licensed Products in the Field in the Territory;

(ii) a non-exclusive license, with the right to grant sublicenses through multiple tiers in accordance with Section 2.2, to the Non-Exclusive Licensed Technology to Exploit the Licensed Compound and Licensed Products in the Field in the Territory and for no other purpose; and

(iii) a non-exclusive license and right of reference, with the right to grant sublicenses through multiple tiers and further rights of reference in accordance with Section 2.2, to the MedImmune Regulatory Documentation that MedImmune or its Affiliates Control as of the Effective Date as necessary for purposes of Exploiting the Licensed Compound and Licensed Products in the Field in the Territory; and

(iv) an exclusive option during the Term of this Agreement to add to the Exclusive Licensed Patents the Licensed Formulation Patents (the "**Option**"). Licensee may exercise the Option, without any additional consideration, up to [***] upon providing written notice to MedImmune. Upon MedImmune's receipt of such written notice, MedImmune and Licensee shall cooperate with each other to take steps to file, if it has not already been done, a portfolio of Licensed Formulation Patents from the Non-Exclusive Licensed Patents that claim priority to U.S. patent application serial no. [***]. Such steps may include, for example, filing separate national and regional phase applications and/or divisional applications claiming priority directly or indirectly to U.S. patent application serial no. [***] relating to the Licensed Compound [***]. MedImmune shall consider in good faith the requests and suggestions of Licensee with respect to filing the portfolio of Licensed Formulation Patents; *provided, however*, that Licensee shall have the right to select in which jurisdictions in the Territory the Licensed Formulation Patents shall

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be filed. Upon Licensee's exercise of the Option, (a) all Licensed Formulation Patents shall be deemed Exclusive Licensed Patents, and (b) MedImmune and its Affiliates shall not prepare, file or prosecute patent claims that specifically relate to, or otherwise recite, the Licensed Compound in any Patents that claim priority directly or indirectly to U.S. patent application serial no. [***].

2.2. Sublicenses. Licensee is permitted to grant sublicenses under the licenses and rights of reference granted in Section 2.1, *provided* that any such sublicenses shall be (i) subject to MedImmune's prior written consent and the prior written consent of any applicable Third Party licensor as required under any In-License Agreement, and (ii) consistent with, and expressly made subject to, the terms and conditions of this Agreement and the In-License Agreements. Licensee shall cause each Sublicensee to comply with the applicable terms and conditions of this Agreement and the In-License Agreements, as if such Sublicensee were a Party to this Agreement. Licensee hereby (x) guarantees the performance of its Affiliates and permitted Sublicensees that are sublicensed as permitted herein and the grant of any such sublicense shall not relieve Licensee of its obligations under this Agreement, except to the extent they are satisfactorily performed by such Sublicensee and (y) waives any requirement that MedImmune exhaust any right, power or remedy, or proceed against any Sublicensee for any obligation or performance under this Agreement prior to proceeding directly against Licensee. Licensee will share such sublicense agreement with MedImmune within [***] days after its execution; *provided* that the financial terms of any such sublicense agreement to the extent not pertinent to an understanding of a Party's obligations or benefits under this Agreement may be redacted.

2.3. Retention of Rights; Limitations Applicable to License Grants.

2.3.1. Retained Rights of MedImmune. Notwithstanding anything to the contrary in this Agreement and without limitation of any rights granted or reserved to MedImmune pursuant to any other term or condition of this Agreement, MedImmune hereby expressly retains, on behalf of itself and its Affiliates (and on behalf of its licensors, (sub)licensees and contractors) all right, title and interest in and to the Excluded Technology.

2.3.2. In-License Agreements. The licenses granted by MedImmune in Section 2.1 include sublicenses under the applicable license rights granted to MedImmune by Third Parties under the In-License Agreements, subject to this Section 2.3.2. Any sublicense with respect to Information or other Intellectual Property of a Third Party hereunder and any right of Licensee (if any) to grant a further sublicense thereunder, shall be subject and subordinate to the terms and conditions of the applicable In-License Agreement, under which such sublicense is granted and shall be effective solely to the extent permitted under the terms of such In-License Agreement. Without limitation of the foregoing, in the event and to the extent that any In-License Agreement requires that particular terms or conditions of such In-License Agreement be contained or incorporated in any agreement

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granting a sublicense thereunder, such terms and conditions are hereby deemed to be incorporated herein by reference and made applicable to the sublicense granted herein under such In-License Agreement.

2.3.3. No Other Rights Granted by MedImmune. Except as expressly provided herein and without limiting the foregoing, MedImmune grants no other right or license.

2.3.4. No Other Rights Granted by Licensee. Except as expressly provided herein, Licensee grants no other right or license to any other Patent, Trademark or other Intellectual Property not otherwise expressly granted herein.

2.4. Licensed Patent Rights Transfer. Promptly (and in no event later than [***] days) following the Effective Date), MedImmune or its designated attorneys, shall provide Licensee, or Licensee's designated attorneys, with its or its attorneys case files for the Licensed Patents . MedImmune shall promptly, upon receipt, forward to Licensee, or Licensee's designated attorneys, any office actions, communications, and correspondence related to the Licensed Patents received by MedImmune after the Effective Date. Additionally, MedImmune shall, from time to time, take such actions as are reasonably requested by Licensee to perfect the license of MedImmune's right, title and interest in the Licensed Patents to Licensee.

2.5. Maintenance of In-License Agreements. MedImmune agrees that it will not, and will ensure that its Affiliates do not, without Licensee's prior written consent (i) sell, assign, transfer, convey, deliver or otherwise divest its interests in any of the In-License Agreements to a Third Party, (ii) mortgage or otherwise encumber its interests in any of the In-License Agreements, in a manner that significantly adversely affects, or would reasonably be expected to significantly adversely affect, Licensee's rights or obligations under this Agreement, (iii) amend any of the In-License Agreements in a manner that significantly adversely affects the rights granted to Licensee under this Agreement, or (iv) undertake any action that would constitute a material breach of, and allow the Third Party that is a party to any In-License Agreements to terminate, any In-License Agreements.

2.6. Completeness of Technology. MedImmune agrees that, if at any time after the Effective Date, MedImmune becomes aware as a result of written notice from Licensee and determines that any Intellectual Property, Inventory or Regulatory Documentation that was owned or Controlled by MedImmune as of the Effective Date and used by MedImmune in the Exploitation of the Licensed Compound or Licensed Products as it existed as of the Effective Date was not included in the Licensed Technology or MedImmune Regulatory Documentation, as applicable, was not included in the license grant to Licensee in Section 2.1 and is deemed necessary for, or was used in, the Exploitation of the Licensed Patents, then MedImmune shall promptly notify Licensee of such determination. MedImmune shall promptly take such actions as may be reasonably necessary to license, provide a right of

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reference to or otherwise transfer such Intellectual Property, Inventory or Regulatory Documentation to Licensee as necessary, in a manner consistent with Sections 2.1 and 3.1, as applicable.

2.7. Exclusivity. MedImmune shall not, and shall not permit any of its Affiliates to, distribute, market, promote, offer for sale or sell the Licensed Compound or Licensed Products directly or indirectly (i) to any Person for commercial use in the Territory. If MedImmune or any of its Affiliates receives or becomes aware of the receipt by a (sub)licensee or distributor of any orders for any Licensed Compound or Licensed Product for commercial use in the Territory, such Person shall refer such orders to Licensee. MedImmune shall cause its Affiliates to notify Licensee of any receipt of any orders for any Licensed Compound or Licensed Product for commercial use in the Territory.

ARTICLE 3
PURCHASE AND SALE OF ASSETS

3.1. Purchase and Sale of Assets. Upon the terms and subject to the conditions set forth in this Agreement, MedImmune hereby sells, conveys, assigns, transfers and delivers to, and shall cause its Affiliates to sell, convey, assign, transfer and deliver to, Licensee, and Licensee hereby purchases and acquires from each of MedImmune or its Affiliates, as the case may be, all of MedImmune's and its Affiliates' right, title and interest in and to the assets described or set forth on Schedule 3.1 attached hereto, including without limitation the Inventory and Product Regulatory Documentation (collectively, the "**Purchased Assets**").

3.2. Excluded Assets. Notwithstanding the provisions of Section 3.1, but subject to Section 2.6, no right, title or interest is being sold, assigned, transferred, conveyed or delivered to Licensee in or to (i) any of the property and assets of MedImmune that are not listed on Schedule 3.1 (or licensed pursuant to Article 2 above), or (ii) any rights or claims of MedImmune under this Agreement (collectively, the "**Excluded Assets**").

3.3. Assumed Liabilities. Subject to the terms and conditions of this Agreement, on and after the Effective Date, Licensee shall assume and agree to pay, perform and discharge all Liabilities and obligations resulting from the ownership and Exploitation of any Purchased Assets or Licensed Product by Licensee to the extent that such Liability arises from any event, condition or circumstance occurring after the Effective Date and not resulting from (i) any breach by MedImmune (or its Affiliates) of any of its obligations under this Agreement, or (ii) MedImmune's (or its Affiliates) gross negligence or willful misconduct (collectively, the "**Assumed Liabilities**").

3.4. Excluded Liabilities. MedImmune shall retain, and shall be responsible for paying, performing and discharging when due, and Licensee shall not assume or have any

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responsibility for paying, performing or discharging, any Liabilities of MedImmune and its Affiliates other than the Assumed Liabilities (collectively, the "**Excluded Liabilities**"). Without limiting the foregoing, neither Licensee nor its Affiliates shall be obligated to assume, and neither of them does assume, and each of them hereby disclaims responsibility for, any of the following Liabilities of MedImmune and its Affiliates:

(i) any Liability attributable to any tangible asset, property or right that is not included in the Purchased Assets;

(ii) any Liability attributable to the research, development or other activity conducted by MedImmune or any Affiliate related to the Product on or prior to the Effective Date; and

(iii) any and all Taxes imposed on the Purchased Assets or that otherwise arise with respect to the use of the Purchased Assets, in each case, for any taxable period (or portion thereof) ending on or prior to the Effective Date, and all Taxes of MedImmune or any of its Affiliates that are or may become payable with respect to all taxable periods, including any Liability for such Taxes that arise as a result of the transactions contemplated by this Agreement.

ARTICLE 4
DEVELOPMENT, REGULATORY AND COMMERCIALIZATION ACTIVITIES; TECHNOLOGY TRANSFER

4.1. Development.

4.1.1. Diligence. After the Effective Date, subject to the Retained Rights, as between the Parties, Licensee shall be solely responsible for all aspects of the Development of the Licensed Compound and Licensed Products in the Field in the Territory at Licensee's own cost and expense, including with respect to any clinical studies or other tests or studies necessary or useful to support the use of a Licensed Product. Without limitation of Section 4.1.2, Licensee shall use Commercially Reasonable Efforts to Develop, and obtain and maintain Regulatory Approvals for, Licensed Products for use in the Field.

4.1.2. Development Plan.

(i) Licensee will share its initial Development Plan and subsequent Development Plans with Medimmune on an annual basis. Licensee will deliver to MedImmune each annual Development Plan not later than [***] days after the end of the Calendar Year.

(ii) Without limitation of Section 4.1.1, Licensee shall perform the Development activities under the current Development Plan and shall use Commercially

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Reasonable Efforts to do so in accordance with the timelines set out in the then-current Development Plan. Licensee shall perform or cause to be performed its Development activities hereunder in good scientific manner and in compliance with Applicable Laws.

4.1.3. Development Costs. Licensee shall be responsible for all of its costs and expenses in connection with the Development of, and obtaining and maintaining Regulatory Approvals for, the Licensed Products in the Field in the Territory.

4.1.4. Development Records. Licensee shall, and shall cause its Affiliates and its and their Sublicensees to, maintain, in good scientific manner, complete and accurate books and records pertaining to Development of Licensed Products hereunder, in sufficient detail to verify compliance with its obligations under this Agreement. Such books and records shall (i) be appropriate for patent and regulatory purposes, (ii) be in compliance with Applicable Law, (iii) properly reflect the work done and results achieved in the performance of its Development activities hereunder, (iv) record only such activities and not include or be commingled with records of activities outside the scope of this Agreement, and (v) be retained by Licensee for at least [***] years after the expiration or termination of this Agreement in its entirety or for such longer period as may be required by Applicable Law. Such books and records shall be subject to MedImmune's audit rights set forth in Section 5.9.

4.1.5. Development Reports. Without limiting Section 4.1.4, at the end of each Calendar Year during which Licensee is conducting Development activities hereunder, Licensee shall provide MedImmune with a summary written report of such Development activities it has performed, or caused to be performed, since the preceding report. Each such summary report shall contain sufficient detail to enable MedImmune to reasonably assess Licensee's compliance with its obligations set out in Sections 4.1.1 and 4.1.2, including: (i) Licensee's, or its Affiliates' or its or their Sublicensees' activities with respect to achieving Regulatory Approvals of Licensed Products and (ii) summaries of clinical study results and other Development activities.

4.2. Regulatory Activities.

4.2.1. Regulatory Approvals. Subject to the Retained Rights, Licensee shall, at its own cost and expense, have the sole right to prepare, obtain and maintain Biologics License Applications (including the setting of the overall regulatory strategy therefor), other Regulatory Approvals and other submissions (including INDs) and to conduct communications with the Regulatory Authorities, for Licensed Products in the Field in the Territory in its name.

4.2.2. Global Safety Database. Licensee shall establish, hold and maintain (at Licensee's sole cost and expense) the global safety database for Licensed Products. MedImmune shall transfer to Licensee the global safety database for the Licensed

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Compound and Licensed Products in existence as of the Effective Date and all Information in the possession and Control of MedImmune as necessary for Licensee to comply with its pharmacovigilance responsibilities in the Territory, including, as applicable, any adverse drug experiences (including those events or experiences that are required to be reported to the FDA under 21 C.F.R. sections 312.32 or 314.80 or to foreign Regulatory Authorities under corresponding Applicable Law outside the United States), from pre-clinical or clinical laboratory, animal toxicology and pharmacology studies, clinical studies and commercial experiences with a Licensed Product, in each case, in the form reasonably requested by Licensee.

4.3. Commercialization.

4.3.1. Diligence. As between the Parties, Licensee shall be solely responsible for Commercialization of the Licensed Products in the Field throughout the Territory at Licensee's own cost and expense. Without limitation of Section 4.3.2, Licensee shall use Commercially Reasonable Efforts to Commercialize the Licensed Products.

4.3.2. Commercialization Plan.

(i) The initial Commercialization of the Licensed Products in the Field in the Territory shall be conducted pursuant to a comprehensive, [***] plan (the "**Commercialization Plan**"). At least [***] days prior to the anticipated date of the First Commercial Sale of any Licensed Product in the first country in the Territory, Licensee shall send to MedImmune the Commercialization Plan. The Commercialization Plan shall include, with respect to the Territory: (a) the general strategies for the promoting, marketing and distributing the Licensed Products; (b) pre-launch Commercialization activities and the expected date of launch; (c) the nature of promotional activities anticipated; (d) non-binding summary-level market and sales forecasts for the Licensed Products; (e) a non-binding projection of Net Sales for Licensed Products; (f) plans regarding distribution and supply chain management; and (g) reimbursement and pricing information.

(ii) Without limitation of Section 4.3.1, Licensee shall perform the Commercialization activities under the applicable Commercialization Plan and shall use Commercially Reasonable Efforts to do so in accordance with the timelines and so as to achieve the objectives set out in the Commercialization Plan.

4.3.3. Commercialization Costs; Booking of Sales; Distribution. Licensee shall be responsible for all of its costs and expenses in connection with the Commercialization of the Licensed Products in the Field in the Territory. Licensee shall invoice and book sales, establish all terms of sale (including pricing and discounts) and warehouse and distribute the Licensed Products in the Field in the Territory and perform or cause to be performed all related services. Licensee shall handle all returns, recalls or

withdrawals, order processing, invoicing, collection, distribution and inventory management with respect to the Licensed Products in the Territory.

4.3.4. Commercialization Records. Licensee shall maintain complete and accurate books and records pertaining to Commercialization of Licensed Products hereunder, in sufficient detail to verify compliance with its obligations under this Agreement and which shall be in compliance with Applicable Law and properly reflect all work done and results achieved in the performance of its Commercialization activities. Such records shall be retained by Licensee for at least [***] years after the expiration or termination of this Agreement in its entirety or for such longer period as may be required by Applicable Law. Such books and records shall be subject to MedImmune's audit rights set forth in Section 5.9.

4.3.5. Commercialization Reports. Without limiting Section 4.3.4, within [***] days following the end of each Calendar Year during which Licensee is conducting Commercialization activities hereunder, Licensee shall provide to MedImmune with written summaries reporting of such Commercialization activities it has performed, or caused to be performed, since the preceding report. Each such report shall contain sufficient detail to enable MedImmune to assess Licensee's compliance with its obligations set out in Sections 4.3.1 and 4.3.2, including in each case: [***].

4.4. Statements and Compliance with Applicable Law. Licensee shall, and shall cause its Affiliates to, comply with all Applicable Laws with respect to the Exploitation of Licensed Products. Without limitation to the foregoing, Licensee shall in all material respects, conform its practices and procedures relating to the Commercialization of the Licensed Products and educating the medical community in the Territory with respect to the Licensed Products to any applicable industry association regulations, policies and guidelines, as the same may be amended from time to time, and Applicable Law.

4.5. Manufacturing. As between the Parties, Licensee shall have the sole responsibility for, at its expense, Manufacturing (or having Manufactured) and supplying the Licensed Compound and Licensed Products for its Development and Commercialization activities in the Territory.

4.6. Subcontracting. Licensee may subcontract with a Third Party to perform any or all of its obligations hereunder (including by appointing one or more distributors); *provided*, that (i) no such permitted subcontracting shall relieve Licensee of obligations hereunder (except to the extent satisfactorily performed by such subcontractor) or any liability and Licensee shall be and remain fully responsible and liable therefor, and (ii) the agreement pursuant to which Licensee engages any Third Party subcontractor must (a) be consistent in all material respects with this Agreement, (b) contain terms obligating such subcontractor to consistent with this Agreement in order to comply with the confidentiality, intellectual property and all other relevant provisions of this Agreement, and (c) contain

terms obligating such subcontractor to permit MedImmune rights of inspection, access and audit substantially similar to those provided to MedImmune in this Agreement. Licensee hereby waives any requirement that MedImmune exhaust any right, power or remedy, or proceed against any subcontractor for any obligation or performance under this Agreement prior to proceeding directly against Licensee.

4.7. Technology Transfer.

4.7.1. To enable Licensee to exercise the rights granted under this Agreement, MedImmune will promptly deliver or otherwise provide to Licensee and Licensee Representatives, Licensed Know-How and the Purchased Assets within the possession or Control of MedImmune or any of its Affiliates. Without limiting the generality of the foregoing, and without limiting the delivery and provision of Licensed Know-How, MedImmune will promptly deliver, make available or otherwise provide to Licensee and Licensee Representatives, Licensed Know-How and the Purchased Assets in accordance with the requirements and timelines set forth in the Transition Activities. Additionally, on a commercially reasonable schedule and in a commercially reasonable format to be agreed upon by the parties, MedImmune will deliver to Licensee or Licensee Representatives, any and all assays, documents, files, diagrams, specifications, designs, schematics, reports, records, data, results, publications, materials, prototypes, test devices, models and simulations, or other written, graphic, biologic, or other tangible material in MedImmune's or its Affiliates' possession in any media, to the extent it discloses or embodies Licensed Know-How. Licensee acknowledges that (i) any materials comprising Inventory transferred by MedImmune to Licensee under this Agreement are experimental in nature and may have unknown characteristics and therefore agrees to use prudence and reasonable care in the use, handling, storage, transportation and disposition and containment of any such materials, and (ii) if Licensee chooses to use such materials in any human application, including in the conduct of any clinical trial, it shall do so at its own risk.

4.7.2. To the extent reasonably requested by Licensee (and for clarity, in addition to the timelines set forth in Transition Activities), MedImmune shall provide reasonable consulting support to Licensee and Licensee Representatives in connection with its Exploitation of Products. MedImmune agrees to use reasonable efforts to (i) make its employees, agents and consultants reasonably available to Licensee (or to Licensee's authorized attorneys and Licensee Representatives) in order for Licensee to fully Exploit the Licensed Products, and (ii) provide contact information in MedImmune's possession and control with respect to the listed inventors of the Licensed Patents, to the extent, in any case, reasonably necessary to enable Licensee (or to Licensee's authorized attorneys and Licensee Representatives) to undertake preparation of U.S. and foreign applications claiming priority to Licensed Patents and prosecution and maintenance of such applications. MedImmune shall only be obligated to complete one (1) technology transfer of all Licensed Know How related to the Manufacture of the Licensed Compound to Licensee or Licensee

Representatives, provided that such technology transfer is complete, as mutually agreed by the Parties, in accordance with the Transition Services Agreement.

4.7.3. In addition to the above, MedImmune will provide support to Licensee, as requested by Licensee, to (i) extend the expiration of dating for any Inventory (including without limitation, Licensed Compound drug substance as well as placebo), (ii) assist (including through Third Party service providers) with filling any Licensed Compound drug substance (and placebo) that is part of the Inventory, (iii) provide on-going stability testing for any such Inventory until the expiration of such Inventory (as extended), (iv) store such Inventory as requested by Licensee to the extent necessary to provide such stability testing, (v) provide other services in order to release Licensed Product drug product that was filled by MedImmune or Third Party service providers, as set out more fully in the Transition Activities.

4.7.4. In order to enable the seamless transfer of technology to Licensee (and/or Licensee Representatives), MedImmune will promptly provide letters of authorization to its Third Party service providers (as requested by Licensee), who have participated in Developing or Manufacturing the Licensed Compound in order to clarify that as of the Effective Date, Licensee has in-licensed and otherwise acquired the rights to the Licensed Compound and Licensed Technology.

4.7.5. Without limiting the foregoing, each Party shall execute and deliver such other instruments and do and perform such other acts and things as reasonably necessary for effecting completely the consummation of the transactions contemplated by this Agreement.

4.8. Cost of MedImmune Support. The Parties hereby agree that the Transition Activities shall be provided at MedImmune's sole expense for up to [***] hours as set out in Schedule 1.104 and/or theTransition Services Agreement, and thereafter at Licensee's sole expense (including MedImmune's employee costs at the FTE Rate).

4.9. Disclosure of Excluded Technology. MedImmune agrees that in the event Licensee is required to disclose Excluded Technology to a Regulatory Authority to obtain Regulatory Approval or otherwise by Applicable Law, it will disclose the required information:

(i) to the Licensee Representatives who have (a) a bona fide need to know such Excluded Technology (for example, those who are directly involved with the preparation, filing and maintenance of any such Regulatory Approval covering the Manufacture and Commercialization of a Product); and (b) who have been fully informed of the highly sensitive nature of the information and the need to maintain its secrecy and avoid inappropriate use. Licensee further agrees to implement procedures via the use of a firewall

or other appropriate means to limit distribution of the Excluded Technology to those Licensee Representatives as described above.

(ii) via a secure dataroom or portal where such dataroom or portal shall be the exclusive source for such Excluded Technology.

ARTICLE 5
PAYMENTS AND RECORDS

5.1. Upfront Payment. In partial consideration of the rights granted by MedImmune to Licensee hereunder, no later than [***] days following the date that Licensee receives and invoice from MedImmune following the Effective Date, Licensee shall pay MedImmune a nonrefundable and noncreditable upfront amount equal to Eight Million Dollars ($8,000,000.00).

5.2. Milestones

5.2.1. Development and Regulatory Milestones. Licensee shall inform MedImmune within [***] Business Days after the occurrence of each of the milestone events listed below. In partial consideration of the rights granted by MedImmune to Licensee hereunder, Licensee shall pay to MedImmune the following one-time payments within [***] days after receipt of an invoice for the achievement of each of the following milestone events which shall be nonrefundable, noncreditable and fully earned upon the achievement of the applicable milestone event:

(i) [***];

(ii) [***];

(iii) [***];

(iv) [***];

(v) [***];

(vi) [***];

(vii) [***];

(viii) [***];

(ix) [***];

(x) [***]; and

(xi) [***].

Each milestone payment in this Section 5.2.1 shall be payable [***]. If, at any time, with respect to a Licensed Product, the achievement of a milestone described in Section 5.2.1 has occurred with respect to which a payment is due hereunder and any of the preceding milestones in this Section 5.2.1 have not been due or been paid , then each such skipped milestone payment shall become due and payable [***], with respect to which payment is due.

5.2.2. Commercial Milestones. In partial consideration of the rights granted by MedImmune to Licensee hereunder, Licensee shall pay to MedImmune the following payments, which shall be nonrefundable, noncreditable and fully earned upon the achievement of the applicable milestone event:

(i) Licensee shall pay to MedImmune [***] in the event that the [***];

(ii) Licensee shall pay to MedImmune [***] in the event that the [***];

(iii) Licensee shall pay to MedImmune [***] in the event that the [***];

(iv) Licensee shall pay to MedImmune [***] in the event that the [***];

(v) Licensee shall pay to MedImmune [***] in the event that the [***];

(vi) Licensee shall pay to MedImmune [***] in the event that the [***];

(vii) Licensee shall pay to MedImmune [***] in the event that the [***]; and

(viii) Licensee shall pay to MedImmune [***] in the event that [***].

Each such milestone payment shall be due within [***] days of the end of the Calendar Quarter in such Calendar Year in which such milestone was achieved. In the event that in [***]. Each milestone payment in this Section 5.2.2 shall be payable only upon the first achievement of such milestone in a given Calendar Year and no amounts shall be due for subsequent or repeated achievements of such milestone in subsequent Calendar Years.

5.2.3. Determination that Milestones Have Occurred. Licensee shall notify MedImmune promptly of the achievement of each of the events identified as a milestone in Section 5.2.1 or Section 5.2.2. In the event that, notwithstanding the fact that Licensee has not provided MedImmune such a notice, MedImmune believes that any such milestone has been achieved, it shall so notify Licensee in writing and the Parties shall promptly meet and discuss in good faith whether such milestone has been achieved. Any dispute under this Section 5.2.3 regarding whether or not such a milestone has been achieved shall be subject to resolution in accordance with Section 11.5.

5.3. Royalties.

5.3.1. Royalty Rates and Reports. As further consideration for the rights granted to Licensee hereunder, commencing upon the First Commercial Sale of a Licensed Product in the Territory, solely during the Royalty Term the Licensee shall pay to MedImmune a royalty on Net Sales of each Licensed Products in the Territory during each Calendar Year at the following rates:

(i) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year less than [***], a royalty rate of [***];

(ii) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year equal to or greater than [***], a royalty rate of [***];

(iii) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year equal to or greater than [***], a royalty rate of [***];

(iv) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year equal to or greater than [***], a royalty rate of [***];

(v) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year equal to or greater than [***], a royalty rate of [***];

(vi) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year equal to or greater than [***], a royalty rate of [***]; and

(vii) for that portion of aggregate Net Sales of all Licensed Products in the Territory during a Calendar Year equal to or greater than [***] a royalty rate of twenty percent (20%).

5.3.2. Each such royalty payment shall be due within [***] days of the end of the Calendar Quarter in such Calendar Year in which such royalty is being achieved. Licensee shall also provide, at the same time each such payment is made, a report showing:

(a) the Net Sales of each Product; (b) the total amount of deductions from Invoiced Sales to determine Net Sales; (c) the applicable royalty rates for each Product on a country-by-country basis in each country in the Territory after applying any adjustments set forth in Section 5.3.4 below; and (d) a calculation of the amount of royalty due to MedImmune.

5.3.3. Blended Royalty. Licensee acknowledges that (i) the Licensed Know-How including the Information included in the MedImmune Regulatory Documentation licensed to Licensee are proprietary and valuable and that without the Licensed Know-How and such Information, Licensee would not be able to obtain and maintain Regulatory Approvals with respect to the Licensed Products, (ii) such Regulatory Approvals will allow Licensee to obtain and maintain regulatory exclusivity with respect to the Licensed Products in the Field in the Territory, (iii) access to the Licensed Know-How and the rights with respect to the MedImmune Regulatory Documentation have provided Licensee with a competitive advantage in the marketplace beyond the exclusivity afforded by the Licensed Patents and (iv) the milestone payments and royalties set out in Section 5.2 and Sections 5.3, respectively, are, in part, intended to compensate MedImmune for such exclusivity and such competitive advantage. The Parties agree that the royalty rates set out in Section 5.3.1 reflect an efficient and reasonable blended allocation of the value provided by MedImmune to Licensee.

5.3.4. Royalty Reductions.

(i) **Know-How Only Royalty Reduction.** During the Royalty Term for a Licensed Product in a country in the Territory, the royalty rates will be reduced by [***] on a country-by-country basis if there is no Valid Claim of a Patent included in the Licensed Patents that Covers the Licensee's (or its Affiliate's or Sublicensee's, as applicable) manufacture, importation or sale of such Licensed Product in such country.

(ii) **Third Party Royalty Reduction.** During the Royalty Term for a Licensed Product in a country in the Territory, the royalty rates will be reduced by [***] of any commercial milestones and/or royalties paid by Licensee (or by its sublicensees) to Third Parties for the Licensed Product. MedImmune shall be solely responsible for paying any and all royalties payable under the In-License Agreements and the royalties paid to MedImmune hereunder are the only royalties that Licensee is due to pay MedImmune (inclusive of any other party of an In-License Agreement); provided that such royalties shall not be offset pursuant to the Third Party Royalty Reduction described above.

(iii) **Biosimilars.** If a Biosimilar Product is launched in a country and any such Biosimilar Product(s) collectively have more than [***] in such country then the royalty rates for such country will be reduced [***].

(iv) **Maximum Reduction.** The annual total of the royalty rates due to MedImmune set out in (i) to (iii) above will not be reduced in aggregate by more than [***].

5.4. Mode of Payment; Offsets. All payments to MedImmune under this Agreement shall be made by deposit of Dollars in the requisite amount to such bank account as MedImmune may from time to time designate by notice to Licensee. For the purpose of calculating any sums due under, or otherwise reimbursable pursuant to, this Agreement (including the calculation of Net Sales expressed in currencies other than Dollars), Licensee shall use the Currency Conversion Policy. For the purposes of this Section 5.4, Currency Conversion Policy means Medimmune's currency conversion policy as of the Effective Date of this Agreement, which is the booking rate for the current month calculated from the average spot rate for [***] of the last [***] Business Days of the previous month, as such spot rate is taken from Reuters as at 08.30 a.m. GMT on each day. Licensee shall have no right to offset, set off or deduct any amounts from or against the amounts due to MedImmune hereunder. The Parties may revise the Currency Conversion Policy by mutual written agreement.

5.5. Taxes.

5.5.1. General. The upfront fee, technology transfer fee, milestone payments and royalties payable by Licensee to MedImmune pursuant to this Agreement (each, a "**Payment**") shall be paid free and clear of any and all Taxes (which, for clarity, shall be the responsibility of Licensee), except for any withholding Taxes required by Applicable Law. Except as provided in this Section 5.5, MedImmune shall be solely responsible for paying any and all Taxes (other than withholding Taxes required by Applicable Law to be deducted from Payments and remitted by Licensee) levied on account of, or measured in whole or in part by reference to, any Payments it receives. Licensee shall deduct or withhold from the Payments any taxes that it is required by Applicable Law to deduct or withhold. Notwithstanding the foregoing, if MedImmune is entitled under any applicable tax treaty to a reduction of rate of, or the elimination of, applicable withholding tax, it may deliver to Licensee or the appropriate Governmental Authority (with the assistance of Licensee to the extent that this is reasonably required and is requested in writing) the prescribed forms necessary to reduce the applicable rate of withholding or to relieve Licensee of its obligation to withhold such tax and Licensee shall apply the reduced rate of withholding or dispense with withholding, as the case may be; *provided* that Licensee has received evidence of MedImmune's delivery of all applicable forms (and, if necessary, its receipt of appropriate governmental authorization) at least [***] days prior to the ime that the Payments are due. If, in accordance with the foregoing, Licensee withholds any amount, it shall pay to MedImmune the balance when due, make timely payment to the proper taxing authority of the withheld amount and send to MedImmune proof of such payment within [***] days following such payment.

5.5.2. Value Added Tax. Notwithstanding anything contained in Section 5.5.1, this Section 5.5.2 shall apply with respect to value added tax ("**VAT**"). All Payments are exclusive of VAT. If any VAT is chargeable in respect of any Payments, Licensee shall pay VAT at the applicable rate in respect of any such Payments following the receipt of a VAT invoice in the appropriate form issued by MedImmune in respect of those Payments, such VAT to be payable on the later of the due date of the payment of the Payments to which such VAT relates and [***] days after the receipt by Licensee of the applicable invoice relating to that VAT payment.

5.6. Anti-Tax Evasion.

5.6.1. In this Section 5.6, (i) references to "**committing tax evasion**" means (a) fraudulently or dishonestly failing to pay any amount of tax to the relevant tax authority within any applicable time limit for the payment of such tax without incurring interest and/or penalties; and (b) **fraudulently** or dishonestly claiming any relief, allowance, credit, deduction, exemption or set off in respect of any tax (or relevant to the computation of any income, profits or gains for the purposes of any tax), or any right to or actual repayment of or saving of tax; and (ii) "**tax**" or "**taxation**" means taxes on gross or net income, profits and gains, and all other taxes, levies, duties, imposts, charges and withholdings of any nature, including any excise, property, wealth, capital, value added, sales, use, occupation, transfer, franchise and payroll taxes and any national insurance or social security contributions, together with all penalties, charges, fees and interest relating to any of the foregoing or to any late or incorrect return in respect of any of them.

5.6.2. The Licensee undertakes that (i) neither it nor its Affiliates shall commit tax evasion; and (ii) it and its Affiliates shall maintain reasonable procedures designed to prevent the Licensee, its Affiliates and any of its employees from undertaking any activities which would facilitate or otherwise result in the Licensee or its Affiliates from committing tax evasion.

5.6.3. The Licensee shall promptly report any apparent breach of this Section 5.6 to MedImmune. The Licensee shall reasonably cooperate with any regulator or public authorities in relation to any investigation relating to the matters referred to in this Section 5.6.

5.7. Interest on Late Payments. If any payment due to either Party under this Agreement is not paid when due, then such paying Party shall pay interest thereon (before and after any judgment) at an annual rate (but with interest accruing on a daily basis) of [***] business day of each month, such interest to run from the date on which payment of such sum became due until payment thereof in full together with such interest., such interest to run from the date on which payment of such sum became due until payment thereof in full together with such interest.

5.8. Financial Records. Licensee shall and shall cause its Affiliates and its and their Sublicensees to, keep true and accurate financial books and records pertaining to the Commercialization of Licensed Products hereunder, including books and records of Invoiced Sales and Net Sales of Licensed Products, in sufficient detail to calculate and verify all amounts payable hereunder. Licensee shall and shall cause its Affiliates and its and their Sublicensees to, retain such books and records until the later of (i) [***] years after the end of the period to which such books and records pertain, (ii) the expiration of the applicable tax statute of limitations (or any extensions thereof), and (iii) for such period as may be required by Applicable Law. for such period as may be required by Applicable Law.

5.9. Audit. At the request of MedImmune, Licensee shall and shall cause its Affiliates and its and their Sublicensees to, permit MedImmune or an independent auditor designated by MedImmune and reasonably acceptable to Licensee, at reasonable times and upon reasonable advance written notice (not less than [***] days), to audit the books and records maintained pursuant to this Agreement, to ensure the accuracy of all reports and payments made hereunder. Such audit right shall not be exercised by MedImmune more than once in any Calendar Year or more than once with respect to sales of a particular Licensed Product in a particular period. All records made available for audit shall be deemed to be Confidential Information of Licensee. Except as provided below the cost of this audit will be borne by MedImmune, unless the audit reveals, with respect to a period, a variance of more than [***] from the reported amounts for such period, in which case Licensee shall bear the cost of the audit. Unless disputed pursuant to Section 5.10 below, and such audit concludes that, (i) additional amounts were owed by Licensee, Licensee shall pay the additional amounts, with interest from the date originally due, or (ii) excess payments were made by Licensee, MedImmune shall reimburse such excess payments; in either case (i) or (ii), within [***] days after the date on which such audit is completed by MedImmune.

5.10. Audit Dispute. The results of each audit, if any, shall be binding on both Parties absent manifest error. In the event of a dispute with respect to any audit under Section 5.9, MedImmune and Licensee shall work in good faith to resolve the disagreement. If the Parties are unable to reach a mutually acceptable resolution of any such dispute within [***] days, the dispute shall be submitted for resolution to a certified public accounting firm jointly selected by each Party's certified public accountants or to such other Person as the Parties shall mutually agree (the "**Auditor**"). The decision of the Auditor shall be final and the costs of such arbitration as well as the initial audit shall be borne between the Parties in such manner as the Auditor shall determine. Not later than [***] days after such decision and in accordance with such decision, Licensee shall pay the additional amounts, with interest from the date originally due, MedImmune shall reimburse the excess payments, as applicable. shall reimburse the excess payments, as applicable.

ARTICLE 6
INTELLECTUAL PROPERTY

6.1. Ownership of Intellectual Property.

6.1.1. Ownership of Technology. As between the Parties, each Party shall own and retain all right, title and interest in and to any and all: (i) Information, and other Intellectual Property that is conceived, discovered, developed or otherwise made by or on behalf of such Party or its Affiliates or its or their (sub)licensees (or Sublicensee(s)), as applicable, under or in connection with this Agreement, whether or not patented or patentable and any and all Patents with respect thereto; and (ii) other Information, inventions, Patents and other Intellectual Property that are owned or otherwise controlled (other than pursuant to the license grants set out in Section 3.1 by such Party or its Affiliates or its or their (sub)licensees (or Sublicensees) (as applicable) outside of this Agreement.

6.1.2. Ownership of Product Trademarks. As between the Parties, Licensee shall own all right, title and interest to the Product Trademarks in the Territory.

6.2. Maintenance and Prosecution of Patents.

6.2.1. In General. As between the Parties, Licensee shall have the right and assume responsibility for, through counsel of its choice, to prepare, file, prosecute and maintain the Exclusive Licensed Patents including directing any related interference, re-issuance, re-examination and opposition proceedings with respect thereto, in each case, at the sole cost and expense of Licensee. If Licensee decides not to prepare, file, prosecute or maintain an Exclusive Licensed Patent in a country in the Territory, Licensee shall provide reasonable prior written notice to MedImmune of such intention and, MedImmune shall thereupon have the right, in its sole discretion, to abandon such Exclusive Licensed Patent or to assume the control and direction of the preparation, filing, prosecution and maintenance of such Exclusive Licensed Patent at MedImmune's sole cost and expense in such country. For the avoidance of doubt, MedImmune shall be responsible for any and all costs associated with the preparation, filing, prosecution and maintenance of Non-Exclusive Licensed Patents.

6.2.2. Prosecuting Party. For purposes of this Section 6.2.2, the Party prosecuting, maintaining or undertaking other related activities with respect to a Patent shall be the "**Prosecuting Party**." The Prosecuting Party shall inform the other Party of all material steps with regard to the preparation, filing, prosecution and maintenance of the Exclusive Licensed Patents, by providing the non-Prosecuting Party with a copy of material communications to and from any patent authority in the Territory regarding such Patents and by providing the non-Prosecuting Party drafts of any material filings or responses to be made to such patent authorities in the Territory sufficiently in advance of submitting such filings or responses so as to allow for a reasonable opportunity for the non-Prosecuting Party to

review and comment thereon. The Prosecuting Party shall consider in good faith the requests and suggestions of the non-Prosecuting Party with respect to such drafts and with respect to strategies for filing and prosecuting such Patents in the Territory.

6.2.3. Cooperation. The non-Prosecuting Party shall, and shall cause its Affiliates to, assist and cooperate with the Prosecuting Party, as the Prosecuting Party may reasonably request from time to time, in the preparation, filing, prosecution and maintenance of the Exclusive Licensed Patents in the Territory under this Agreement, including that the non-Prosecuting Party shall, and shall reasonably endeavor to ensure that its Affiliates (i) offer its comments, if any, promptly, and (ii) provide access to relevant documents and other evidence and make its employees available at reasonable business hours; *provided*, *however*, that the Prosecuting Party shall reimburse the non-Prosecuting Party for its reasonable and verifiable out-of-pocket costs and expenses incurred in connection therewith.

6.2.4. Patent Term Extension and Supplementary Protection Certificate. As between the Parties, Licensee shall have the sole right to make decisions regarding and to apply for, patent term extensions, in the Territory, including in the United States with respect to extensions pursuant to 35 U.S.C. §156 et. seq. and in other jurisdictions pursuant to supplementary protection certificates, and in all jurisdictions with respect to any other extensions that are now or become available in the future, wherever applicable, for the Exclusive Licensed Patents and the Licensed Products, in each case including whether or not to do so; *provided* that MedImmune shall consult with Licensee to determine the course of action with respect to such filings. MedImmune shall provide prompt and reasonable assistance, as requested by Licensee, including by taking such reasonable action as patent holder as is required under any Applicable Law to obtain such extension or supplementary protection certificate.

6.2.5. Patent Listings. As between the Parties, Licensee shall have the sole right to make decisions regarding and Licensee shall have the right to make all filings with Regulatory Authorities in the Territory with respect to the Exclusive Licensed Patents including as required or allowed (i) in the United States, in the FDA's Purple Book and (ii) in the European Union, under the national implementations of Article 10.1(a)(iii) of Directive 2001/EC/83 or other international equivalents.

6.3. Enforcement of Patents.

6.3.1. Notice. Each Party shall promptly notify the other Party in writing of (i) any alleged or threatened infringement of the Exclusive Licensed Patents, in any jurisdiction in the Territory or (ii) any certification, claim, demand, action or cause of action for declaratory relief alleging that any claim in an Exclusive Licensed Patent is invalid or unenforceable or alleging that any claim of an Exclusive Licensed Patent would not be infringed by the making, use, offer for sale, sale or import of a product or any equivalent or

similar certification or notice in any other jurisdiction in the Territory, in each case ((i) and (ii)) of which such Party becomes aware (an "**Infringement**").

6.3.2. Enforcement of Patents. As between the Parties, (i) Prosecuting Party pursuant to 6.2.2 shall have the first right, but not the obligation, to prosecute any Infringement with respect to the Exclusive Licensed Patents including as a defense or counterclaim in connection with any Third Party Infringement Claim, at Prosecuting Party's sole cost and expense, using counsel of Prosecuting Party's choice and (ii) MedImmune shall have the sole right, but not the obligation, to prosecute Infringement with respect to the Non-Exclusive Licensed Technology, including as a defense or counterclaim in connection with any Third Party Infringement Claim, at MedImmune's sole cost and expense, using counsel of its choice. For purposes of this Section 6.3, the Party prosecuting any Infringement pursuant to the foregoing sentence with respect to a Patent shall be the "**Enforcing Party**." In the event MedImmune prosecutes any such Infringement in the Field in the Territory, Licensee shall have the right to join as a party to such claim, suit or proceeding and participate with its own counsel at its sole cost and expense; *provided* that MedImmune shall retain control of the prosecution of such claim, suit or proceeding, including the response to any defense or defense of any counterclaim raised in connection therewith. In the event Licensee prosecutes any such Infringement in the Field in the Territory, MedImmune shall have the right to join as a party to such claim, suit or proceeding and participate with its own counsel at its sole cost and expense; *provided* that Licensee shall retain control of the prosecution of such claim, suit or proceeding, including the response to any defense or defense of any counterclaim raised in connection therewith. If the Enforcing Party or its designee does not take commercially reasonable steps to prosecute an Infringement in the Field (x) within [***] days following the first notice provided above with respect to such Infringement or (y) *provided* such date occurs after the first such notice of such Infringement is provided, [***] Business Days before the time limit, if any, set out in appropriate laws and regulations for filing of such actions, whichever comes first, then (1) the Enforcing Party shall so notify the non-Enforcing Party and (2) subject to any rights of any Third Parties under any In-License Agreements (or other applicable Third Party agreements existing as of the Effective Date) and upon the Enforcing Party's written consent (such consent not to be unreasonably withheld, conditioned or delayed), the non-Enforcing may prosecute such alleged or threatened infringement in the Field at its sole cost and expense, whereupon the non-Enforcing Party shall be deemed the Enforcing Party with respect to such Infringement.

6.3.3. Cooperation. The Parties agree to cooperate fully in any Infringement action pursuant to this Section 6.3, including by making the inventors, applicable records and documents (including laboratory notebooks) with respect to the relevant Patents available to the Enforcing Party on the Enforcing Party's request. With respect to an action controlled by the applicable Enforcing Party, the other Party shall, and shall cause its Affiliates to, assist and cooperate with the Enforcing Party, as the Enforcing

Party may reasonably request from time to time, in connection with its activities set out in this Section, including where necessary, furnishing a power of attorney solely for such purpose or joining in, or being named as a necessary party to, such action, providing access to relevant documents and other evidence and making its employees available at reasonable business hours; *provided* that the Enforcing Party shall reimburse such other Party for its reasonable and verifiable out-of-pocket costs and expenses incurred in connection therewith. Unless otherwise set out herein, the Enforcing Party shall have the right to settle such claim; *provided* that neither Party shall have the right to settle any Infringement litigation under this Section 6.3 in a manner that has a material adverse effect on the rights or interest of the other Party or in a manner that imposes any costs or liability on or involves any admission by, the other Party, without the express written consent of such other Party (which consent shall not be unreasonably withheld, conditioned or delayed). In connection with any activities with respect to an Infringement action prosecuted by the applicable Enforcing Party pursuant to this Section 6.3 involving Patents Controlled by or licensed under Article 2 to the other Party, the Enforcing Party shall (i) consult with the other Party as to the strategy for the prosecution of such claim, suit or proceeding, (ii) consider in good faith any comments from the other Party with respect thereto and (iii) keep the other Party reasonably informed of any material steps taken and provide copies of all material documents filed, in connection with such action.

6.3.4. Recovery. Except as otherwise agreed by the Parties in connection with a cost sharing arrangement, any recovery realized as a result of such litigation described above in this Section 6.3 (whether by way of settlement or otherwise) shall be first, allocated to reimburse the Parties for their costs and expenses in making such recovery (which amounts shall be allocated pro rata if insufficient to cover the totality of such expenses). Any remainder after such reimbursement is made shall be [***]; *provided*, *however*, [***].

6.4. Infringement Claims by Third Parties. If the Exploitation of a Licensed Product in the Territory pursuant to this Agreement results in, or is reasonably expected to result in, any claim, suit or proceeding by a Third Party alleging infringement by Licensee or any of its Affiliates or its or their Sublicensees, (a "**Third Party Infringement Claim**"), including any defense or counterclaim in connection with an Infringement action initiated pursuant to Section 6.3.2, the Party first becoming aware of such alleged infringement shall promptly notify the other Party thereof in writing. As between the Parties, Licensee shall be responsible for defending any such claim, suit or proceeding, at its sole cost and expense using counsel of Licensee's choice, in relation to technology licensed under any Exclusive Licensed Technology, and MedImmune shall be responsible for defending any such claim, suit or proceeding at proceeding at its sole cost and expense, using counsel of MedImmune's choice in relation to technology licensed under any Non-Exclusive Licensed Technology. MedImmune shall, and shall cause its Affiliates to, assist and cooperate with Licensee, as Licensee may reasonably request from time to time, in connection with its activities set out in this Section 6.4, including where necessary, furnishing a power of attorney solely for such

purpose or joining in, or being named as a necessary party to, such action, providing access to relevant documents and other evidence and making its employees available at reasonable business hours; *provided* that Licensee shall reimburse MedImmune for its reasonable and verifiable out-of-pocket costs and expenses incurred in connection therewith. Licensee shall keep MedImmune reasonably informed of all material developments in connection with any such claim, suit or proceeding. Licensee agrees to provide MedImmune with copies of all material pleadings filed in such action and to allow MedImmune reasonable opportunity to participate in the defense of the claims. Any damages, or awards, including royalties incurred or awarded in connection with any Third Party Infringement Claim defended under this Section 6.4 shall be [***].

6.5. Invalidity or Unenforceability Defenses or Actions. As between the Parties, (i) Prosecuting Party pursuant to 6.2.2 shall have the first right, but not the obligation, to defend and control the defense of the validity and enforceability of the Exclusive Licensed Patents at its sole cost and expense, using counsel of Prosecuting Party's choice and including when such invalidity or unenforceability is raised as a defense or counterclaim in connection with an Infringement action initiated pursuant to Section 6.3. For purposes of this Section 6.5, the Party prosecuting any Infringement pursuant to the foregoing sentence with respect to a Patent shall be the "**Controlling Party**." With respect to any such claim, suit or proceeding in the Territory, the non-Controlling Party may participate in such claim, suit or proceeding with counsel of its choice at its sole cost and expense; *provided* that the Controlling Party shall retain control of the defense in such claim, suit or proceeding. If the Controlling Party or its designee elects not to defend or control the defense of the applicable Patents in a suit brought in the Territory or otherwise fails to initiate and maintain the defense of any such claim, suit or proceeding, then subject to any rights of Third Parties under any In-License Agreements (or other applicable Third Party agreements existing as of the Effective Date) the non-Controlling Party may conduct and control the defense of any such claim, suit or proceeding at its sole cost and expense. The non-Controlling Party in such an action shall, and shall cause its Affiliates to, assist and cooperate with the Controlling Party, as such Controlling Party may reasonably request from time to time, in connection with its activities set out in this Section 6.5, including where necessary, furnishing a power of attorney solely for such purpose or joining in, or being named as a necessary party to, such action, providing access to relevant documents and other evidence and making its employees available at reasonable business hours; *provided* that the Controlling Party shall reimburse the non-Controlling Party for its reasonable and verifiable out-of-pocket costs and expenses incurred in connection therewith. In connection with any activities with respect to a defense, claim or counterclaim relating to the Exclusive Licensed Patents, pursuant to this Section 6.5, the Controlling Party shall (x) consult with the non-Controlling Party as to the strategy for such activities, (y) consider in good faith any comments from the non-Controlling Party and (z) keep the non-Controlling Party reasonably informed of any material steps taken and provide copies of all material documents filed, in connection with such defense, claim or counterclaim.

6.6. Third Party Patent Rights. Exploitation of the Licensed Compound or Licensed Product in the Field and in the Territory by Licensee, any of its Affiliates or any of its or their Sublicensees infringes or is reasonably expected to infringe any Patent of a Third Party in any country in the Territory (such right, a "**Third Party Patent Right**"), then, as between the Parties, Licensee shall have the first right, but not the obligation, to negotiate and obtain a license from such Third Party to such Third Party Patent Right as necessary or desirable for Licensee or its Affiliates or its or their Sublicensees to Exploit the Licensed Compound and Licensed Products in the Field in such country; *provided* that (i) [***], and (ii) [***]. Licensee shall provide prior notice to MedImmune before entering into any such license and will consult with Medimmune with respect to such matter (subject to Section 7.6 hereof) and consider feedback provided by MedImmune in good faith.

6.7. Product Trademarks.

6.7.1. Notice. Each Party shall provide to the other Party prompt written notice of any actual or threatened infringement of the Product Trademarks in the Territory and of any actual or threatened claim that the use of the Product Trademarks in the Territory violates the rights of any Third Party, in each case, of which such Party becomes aware.

6.7.2. Prosecution of Product Trademarks. Licensee shall be responsible for the registration, prosecution and maintenance of the Product Trademarks using counsel of its own choice. All costs and expenses of registering, prosecuting and maintaining the Product Trademarks shall be borne solely by Licensee.

6.7.3. Enforcement of Product Trademarks. Licensee shall have the right to take such action as Licensee deems necessary against a Third Party based on any alleged, threatened or actual infringement, dilution, misappropriation or other violation of or unfair trade practices or any other like offense relating to, the Product Trademarks by a Third Party in the Territory at its sole cost and expense and using counsel of its own choice. Licensee shall retain any damages or other amounts collected in connection therewith.

6.7.4. Third Party Claims. Licensee shall have the right to defend against any alleged, threatened or actual claim by a Third Party that the use or registration of the Product Trademarks in the Territory infringes, dilutes, misappropriates or otherwise violates any Trademark or other right of that Third Party or constitutes unfair trade practices or any other like offense or any other claims as may be brought by a Third Party against a Party in connection with the use of the Product Trademarks with respect to a Licensed Product in the Territory at its sole cost and expense and using counsel of its choice. Any damages, or awards, including royalties incurred or awarded in connection with any such claim defended under this Section 6.7.4 shall be borne by Licensee.

6.7.5. Cooperation. MedImmune shall, and shall cause its Affiliates to, assist and cooperate with Licensee, as Licensee may reasonably request from time to time, in

connection with its activities set out in this Section, including where necessary, furnishing a power of attorney solely for such purpose or joining in, or being named as a necessary party to, such action, providing access to relevant documents and other evidence and making its employees available at reasonable business hours; *provided* that Licensee shall reimburse MedImmune for its and its Affiliates' reasonable and verifiable out-of-pocket costs and expenses incurred in connection therewith.

ARTICLE 7
CONFIDENTIALITY AND NON-DISCLOSURE

7.1. Confidentiality Obligations. Each Party shall and shall cause its officers, directors, employees, advisors and agents to, keep confidential and not publish or otherwise disclose to a Third Party and not use, directly or indirectly, for any purpose, any Confidential Information furnished or otherwise made known to it, directly or indirectly, by the other Party, except to the extent such disclosure or use is solely for the purpose of exercising its rights and performing its obligations hereunder. "**Confidential Information**" means any technical, business or other information provided by or on behalf of one Party to the other Party in connection with this Agreement, whether prior to, on or after the Effective Date, including information relating to the terms of this Agreement, information relating to the Licensed Compound or any Licensed Product (including the Regulatory Documentation), any Development or Commercialization of the Licensed Compound or any Licensed Product, any know-how with respect thereto developed by or on behalf of the disclosing Party or its Affiliates, or the scientific, regulatory or business affairs or other activities of either Party. For purposes of clarity, on and after the Effective Date, all Confidential Information concerning the Licensed Compound and any Licensed Product transferred to Licensee under this Agreement, shall be considered Confidential Information of Licensee, including any such Confidential Information generated by MedImmune prior to the Effective Date. Notwithstanding the foregoing, the confidentiality and non-use obligations under this Section 7.1 with respect to any Confidential Information shall not include any information that:

(i) is or hereafter becomes part of the public domain by public use, publication, general knowledge or the like through no breach of this Agreement by the receiving Party (or its affiliates);

(ii) can be demonstrated by documentation or other competent proof to have been in the receiving Party's (or its Affiliates) possession prior to disclosure by the disclosing Party without any obligation of confidentiality with respect to such information;

(iii) is subsequently received by the receiving Party (or its Affiliates) from a Third Party who is not bound by any obligation of confidentiality with respect to such information;

(iv) has been published by a Third Party or otherwise enters the public domain through no fault of the receiving Party (or its Affiliates) in breach of this Agreement; or

(v) can be demonstrated by documentation or other competent evidence to have been independently developed by or for the receiving Party (or its Affiliates) without reference to the disclosing Party's Confidential Information.

Specific aspects or details of Confidential Information shall not be deemed to be within the public domain or in the possession of the receiving Party merely because the Confidential Information is embraced by more general information in the public domain or in the possession of the receiving Party. Further, any combination of Confidential Information shall not be considered in the public domain or in the possession of the receiving Party merely because individual elements of such Confidential Information are in the public domain or in the possession of the receiving Party unless the combination and its principles are in the public domain or in the possession of the receiving Party.

7.2. Permitted Disclosures. Each Party may disclose Confidential Information to the extent that such disclosure is:

(i) made in response to a valid order of a court of competent jurisdiction or other supra-national, federal, national, regional, state, provincial and local Governmental Entity or regulatory body of competent jurisdiction or, if in the reasonable opinion of the receiving Party's legal counsel, such disclosure is otherwise required by Applicable Law, including by reason of filing with securities regulators; *provided, however*, that the receiving Party shall first have given notice to the disclosing Party and given the disclosing Party a reasonable opportunity to quash such order or to obtain a protective order or confidential treatment requiring that the Confidential Information and documents that are the subject of such order be held in confidence by such court or agency or, if disclosed, be used only for the purposes for which the order was issued; and provided, further, that the Confidential Information disclosed in response to such court or governmental order shall be limited to that information which is legally required to be disclosed in response to such court or governmental order;

(ii) made by or on behalf of the receiving Party to the Regulatory Authorities as required in connection with any filing, application or request for Regulatory Approval; *provided, however*, that reasonable measures shall be taken to assure confidential treatment of such information to the extent practicable and consistent with Applicable Law; or

(iii) made by or on behalf of the receiving Party to a patent authority as may be reasonably necessary or useful for purposes of obtaining or enforcing a

Patent; provided, however, that reasonable measures shall be taken to assure confidential treatment of such information, to the extent such protection is available.

7.3. Use of Name. The use of name or Trademark of the other Party or any of its Affiliates or any of its or their (sub)licensees (or Sublicensees) (or any abbreviation or adaptation thereof) in any publication, press release, marketing and promotional material or other form of publicity without the prior written approval of such other Party is not permitted. The restrictions imposed by this Section 7.3 do not restrict (i) either Party from making any disclosure identifying the other Party to the extent required in connection with its exercise of its rights or obligations under this Agreement and (ii) either Party from making any disclosure identifying the other Party that is required by Applicable Law or the rules of a stock exchange on which the securities of the disclosing Party are listed (or to which an application for listing has been submitted

7.4. Public Announcements and Non-Disclosure of Agreement.

7.4.1. Restrictions. The Parties agree that neither Party shall (i) disclose the existence or terms of this Agreement or the terms of any term sheet or agreement negotiated pursuant to Section 2.1, or (ii) issue any press release or public statement disclosing information relating to this Agreement or the transactions contemplated hereby or the terms hereof, without prior written consent of the other Party, which consent shall not be unreasonably withheld, conditioned or delayed (or as such consent may be obtained in accordance with Section 7.4.2). Notwithstanding the foregoing, either Party may disclose the existence and terms of this Agreement to its Affiliates, and to its (actual or potential) permitted licensees, sublicensees, acquirers or assignees and subcontractors (and their advisors) and to investment bankers, investors, lenders, accountants and legal advisors and to such Party's directors, employees, contractors and agents, who have a need to know such Confidential Information. Each Party shall advise any such permitted licensees, sublicensees, acquirers or assignees, subcontractors (and their advisors), investment bankers, investors, lenders, accountants and legal advisors and such Party's directors, employees, contractors and agents who receive Confidential Information of the confidentiality obligations set forth in Article 7 for at least [***] years, and such Party shall take steps to ensure (through enforcement of written agreements or otherwise) that they comply with such obligations as if they had been a Party hereto; provided, however, that such Party shall remain responsible for any failure by any Person who receives such information from such Party pursuant to this Section 7.4 to treat such information as required under this Article 7.

7.4.2. Review. In the event either Party (the "**Issuing Party**") is required by Law or the rules or regulations of any applicable United States securities exchange or regulatory or governmental body to which the relevant Party is subject to issue a press release or other public statement disclosing information relating to this Agreement or the transactions contemplated hereby or the terms hereof, the Issuing Party will provide the

other Party (the "**Reviewing Party**") with a copy of the proposed press release or public statement (the "**Release**"). The Issuing Party will specify with each such Release, taking into account the urgency of the matter being disclosed, a reasonable period of time within which the Receiving Party may provide any comments on such Release (but in no event less than [***] Business Days, unless earlier disclosure is required) and if the Receiving Party fails to provide any comments during the response period called for by the Issuing Party, the Reviewing Party will be deemed to have consented to the issuance of such Release. If the Receiving Party provides any comments, the Parties will consult on such Release and work in good faith to prepare a mutually acceptable Release. Either Party may subsequently publicly disclose any information previously contained in any Release so consented to. For the avoidance of doubt, Licensee, in its sole discretion, may disclose the results or status of research, development or any clinical trial conducted by Licensee or any health or safety matter related to any Licensed Compound or Licensed Products.

7.5. Return of Confidential Information. Upon the effective date of the expiration or termination of this Agreement for any reason, either Party may request in writing and the non-requesting Party shall either, with respect to Confidential Information of the requesting Party to which such non-requesting Party does not retain rights under the surviving provisions of this Agreement, at the requesting Party's election, (i) promptly destroy all copies of such Confidential Information in the possession or control of the non-requesting Party and confirm such destruction in writing to the requesting Party or (ii) promptly deliver to the requesting Party, at the non-requesting Party's sole cost and expense, all copies of such Confidential Information in the possession or control of the non-requesting Party. Notwithstanding the foregoing, the non-requesting Party shall be permitted to retain such Confidential Information (x) to the extent necessary or useful for purposes of performing any continuing obligations or exercising any ongoing rights hereunder and, in any event, a single copy of such Confidential Information for archival purposes and (y) any computer records or files containing such Confidential Information that have been created solely by such non-requesting Party's automatic archiving and back-up procedures, to the extent created and retained in a manner consistent with such non-requesting Party's standard archiving and back-up procedures, but not for any other uses or purposes. All Confidential Information shall continue to be subject to the terms of this Agreement until [***].

7.6. Privileged Communications. The Parties may, from time to time, disclose to one another privileged communications with counsel, including opinions, memoranda, letters and other written, electronic and verbal communications. Such disclosures are made with the understanding that they shall remain confidential in accordance with this Article 7, that they will not be deemed to waive any applicable attorney-client or attorney work product or other privilege and that they are made in connection with the shared community of legal interests existing between MedImmune and Licensee, including the community of legal interests in avoiding infringement of any valid, enforceable patents of Third Parties prosecuting and maintaining the validity of the Licensed Patents. In the event of any

litigation (or potential litigation) with a Third Party related to this Agreement or the subject matter hereof, the Parties shall, upon either Party's request, enter into a reasonable and customary joint defense agreement. In any event, each Party shall consult in a timely manner with the other Party before engaging in any conduct (*e.g.*, producing information or documents) in connection with litigation or other proceedings that could conceivably implicate privileges maintained by the other Party. Notwithstanding anything contained in this Section 7.6, nothing in this Agreement shall prejudice a Party's ability to take discovery of the other Party in disputes between them relating to the Agreement and no information otherwise admissible or discoverable by a Party shall become inadmissible or immune from discovery solely by this Section 7.6.

ARTICLE 8
REPRESENTATIONS AND WARRANTIES

8.1. Mutual Representations and Warranties. MedImmune and Licensee each represents and warrants to the other, as of the Effective Date, and covenants, that:

8.1.1. It is a duly organized, validly existing and in good standing under the laws of the jurisdiction of its organization and has all requisite power and authority, corporate or otherwise, to execute, deliver and perform this Agreement;

8.1.2. The execution and delivery of this Agreement and the performance by it of the transactions contemplated hereby have been duly authorized by all necessary corporate action and do not violate: (i) such Party's charter documents, bylaws or other organizational documents; (ii) in any material respect, any agreement, instrument or contractual obligation to which such Party is bound; (iii) any requirement of any Applicable Law; or (iv) any order, writ, judgment, injunction, decree, determination or award of any court or governmental agency presently in effect applicable to such Party;

8.1.3. This Agreement is a legal, valid and binding obligation of such Party enforceable against it in accordance with its terms and conditions, subject to the effects of bankruptcy, insolvency or other laws of general application affecting the enforcement of creditor rights, judicial principles affecting the availability of specific performance and general principles of equity (whether enforceability is considered a proceeding at law or equity);

8.1.4. It is not under any obligation, contractual or otherwise, to any Person that conflicts with or is inconsistent in any material respect with the terms of this Agreement or that would impede the diligent and complete fulfillment of its obligations hereunder; and

8.1.5. Neither Party nor any its or their Affiliates' employees nor, to such Party's Knowledge, any employees of their respective licensees, contractors, agents and

consultants who have been involved, on its behalf, in the Exploitation of the Licensed Product:

(i) is debarred under Section 306(a) or 306(b) of the FFDCA or by the analogous Applicable Laws of any Regulatory Authority;

(ii) has been charged with, or convicted of, any felony or misdemeanor within the ambit of 42 U.S.C. §§ 1320a-7(a), 1320a-7(b)(l)-(3), or pursuant to the analogous Applicable Laws of any Regulatory Authority, or is proposed for exclusion, or the subject of exclusion or debarment proceedings by a Regulatory Authority; or

(iii) is excluded, suspended or debarred from participation, or otherwise ineligible to participate, in any U.S. or non-U.S. healthcare programs (or has been convicted of a criminal offense that falls within the scope of 42 U.S.C. §1320a-7 but not yet excluded, debarred, suspended, or otherwise declared ineligible), or excluded, suspended or debarred by a Regulatory Authority from participation, or otherwise ineligible to participate, in any procurement or non-procurement programs.

8.2. Additional Representations and Warranties of MedImmune. MedImmune further represents and warrants to Licensee, as of the Effective Date, that:

8.2.1. Title to Assets. MedImmune or its Affiliates have good and valid title to all of the Purchased Assets in their entirety free and clear of all encumbrances and liabilities. Schedule 3.1 is a complete list of all Purchased Assets, including the Inventory, as it exists as of the Effective Date.

8.2.2. Litigation and Claims. There is no action, suit, claim, proceeding or investigation pending that has been served on MedImmune, and to the Knowledge of MedImmune, there is no other action, suit, claim, proceeding or investigation pending or threatened against MedImmune before or by any federal, state, municipal or other governmental court, agency or instrumentality, which would prevent MedImmune's performance of this Agreement and the transactions contemplated hereby.

8.2.3. Intellectual Property Rights.

(i) MedImmune has sufficient legal and/or beneficial ownership and/or rights in the Licensed Technology necessary to grant the licenses and sublicenses set forth in accordance with the terms of this Agreement.

(ii) None of the Licensed Technology constitute Third Party Intellectual Property.

(iii) Schedule 1.39 lists and separately identifies each and every Exclusive Licensed Patent. The list of Exclusive Licensed Patents included on Schedule 1.39 is a complete list of all Patents Controlled by MedImmune or its Affiliates prior to the Effective Date that [***] relate to the Licensed Compound or Licensed Products, or the Exploitation thereof, as the Licensed Compound or Licensed Products exist as of the Effective Date.

(iv) MedImmune has diligently prosecuted the Exclusive Licensed Patents, taken all necessary actions and paid all necessary fees to maintain the Licensed Patents in full force and effect, including, to MedImmune's Knowledge, complying with the duty of candor and disclosure to the United States Patent and Trademark Office and any relevant foreign patent office with respect to all Exclusive Licensed Patents and have made no misrepresentation in the Exclusive Licensed Patents. To MedImmune's Knowledge, the Exclusive Licensed Patents are valid, subsisting and enforceable.

(v) Schedule 8.2.3(y) accurately identifies and describes each action, filing and payment that MedImmune is aware of as of the Effective Date that must be taken or made on or before the date that is [***] months after the date of this Agreement to maintain such item of Exclusive Licensed Patents in full force and effect.

(vi) To MedImmune's Knowledge, MedImmune has had and currently has all rights in Exclusive Licensed Patents necessary to carry out MedImmune's former activities and current activities with respect to the Licensed Compound and Licensed Product (including, without limitation, the design, development, use and provision thereof) and there is no other Intellectual Property necessary to carry out MedImmune's business as currently conducted.

(vii) Title to all Exclusive Licensed Patents owned or purported to be owned by MedImmune, whether beneficially or otherwise, is held by and in the name of MedImmune. The transactions contemplated under this Agreement will not alter, impair or otherwise affect any rights of MedImmune in any Exclusive Licensed Patents.

(viii) No actual claims or, to MedImmune's Knowledge, threatened claims challenging the validity, enforceability, effectiveness or ownership by MedImmune of any of the Exclusive Licensed Patents, exist, nor to MedImmune's Knowledge is there any valid basis for such a claim.

(ix) There are no legal proceedings, including without limitation litigation, interference, re-examination, reissue, opposition, nullity, or cancellation proceedings, pending with respect to any of the Exclusive Licensed Patents.

(x) Except as set forth on Schedule 8.2.3(x), to MedImmune's Knowledge, there is no unauthorized use, infringement or misappropriation by any third party or current or former employee of MedImmune of any Exclusive Licensed Patents.

(xi) MedImmune has taken commercially reasonable measures to protect its ownership of, and rights in, all Licensed Technology.

(xii) MedImmune has not granted any licenses or covenants not to sue under the Exclusive Licensed Patents.

(xiii) MedImmune has paid all licensing fees, royalties, profit participations and other payments that were due or payable by MedImmune or any of its Affiliates in connection with its use or practice of the Exclusive Licensed Patents prior to the Effective Date.

(xiv) The In-License Agreements are a complete list of all agreements to which MedImmune is a party granting or assigning any right (whether contingent or otherwise) to own, use or practice any rights under any Licensed Technology.

(xv) All of the In-License Agreements are in force and effect in all material respects as of the date hereof and are binding and enforceable against MedImmune and, to MedImmune's Knowledge, any other party to each such In-License Agreements

(xvi) Except as set forth on Schedule 8.2.3(xvi), (a) neither MedImmune nor, to MedImmune's Knowledge, any other party thereto is in, or has received notice of, material breach or material default of any In-License Agreement, and (b) no event has occurred that with notice or lapse of time would constitute a material breach or material default under any In-License Agreement by MedImmune or, to MedImmune's Knowledge, any other party to any In-License Agreement or would permit the modification or premature termination of any In-License Agreement by any other party thereto.

(xvii) MedImmune has delivered to Licensee, or made available to Licensee or its advisors, copies of each In-License Agreement and included all amendments or modifications thereto.

(xviii) As of the date hereof, MedImmune has not received any written notice from any third party asserting a claim, or threatening to make a claim, which would adversely affect the rights of Licensee as a sublicensee under any In-License Agreement.

8.3. Additional Representations and Warranties of Licensee. As of the Effective Date, Licensee: (i) has conducted its own investigation and analysis of (a) the Patent and other proprietary rights of Third Parties as such rights relate to the Exploitation of

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the Licensed Compound as contemplated hereunder and (b) the potential infringement thereof; (ii) understands the complexity and uncertainties associated with possible claims of Infringement of Patent or other proprietary rights of Third Parties, particularly those relating to pharmaceutical products; and (iii) acknowledges and agrees that it is solely responsible for the risks of such claims after the Effective Date.

8.4. DISCLAIMER OF WARRANTIES. EXCEPT FOR THE REPRESENTATIONS AND WARRANTIES EXPRESSLY SET FORTH IN THIS ARTICLE 8, BOTH PARTIES DISCLAIM ALL OTHER REPRESENTATIONS AND WARRANTIES, WHETHER WRITTEN OR ORAL OR EXPRESS OR IMPLIED, INCLUDING ANY WARRANTY OF QUALITY, MERCHANTABILITY OR FITNESS FOR A PARTICULAR USE OR PURPOSE OR ANY WARRANTY AS TO THE VALIDITY OF ANY PATENTS OR THE NON-INFRINGEMENT OF ANY INTELLECTUAL PROPERTY RIGHTS OF THIRD PARTIES. FITNESS FOR A PARTICULAR USE OR PURPOSE OR ANY WARRANTY AS TO THE VALIDITY OF ANY PATENTS OR THE NON-INFRINGEMENT OF ANY INTELLECTUAL PROPERTY RIGHTS OF THIRD PARTIES.

8.5. ADDITIONAL WAIVER. LICENSEE AGREES THAT: (i) THE LICENSED PATENTS ARE LICENSED "AS IS," "WITH ALL FAULTS," AND "WITH ALL DEFECTS," AND LICENSEE EXPRESSLY WAIVES ALL RIGHTS TO MAKE ANY CLAIM WHATSOEVER AGAINST MEDIMMUNE FOR MISREPRESENTATION OR FOR BREACH OF PROMISE, GUARANTEE OR WARRANTY OF ANY KIND RELATING TO THE LICENSED PATENTS; (ii) LICENSEE AGREES THAT MEDIMMUNE WILL HAVE NO LIABILITY TO LICENSEE FOR ANY ACT OR OMISSION IN THE PREPARATION, FILING, PROSECUTION, MAINTENANCE, ENFORCEMENT, DEFENCE OR OTHER HANDLING OF THE LICENSED PATENTS; AND (iii) LICENSEE IS SOLELY RESPONSIBLE FOR DETERMINING WHETHER THE LICENSED PATENTS HAVE APPLICABILITY OR UTILITY IN LICENSEE'S CONTEMPLATED EXPLOITATION OF THE LICENSED PRODUCTS AND LICENSEE ASSUMES ALL RISK AND LIABILITY IN CONNECTION WITH SUCH DETERMINATION.

8.6. Anti-Bribery and Anti-Corruption Compliance.

8.6.1. Licensee agrees, on behalf of itself, its officers, directors and employees and on behalf of its Affiliates, agents, representatives, consultants and subcontractors hired in connection with the subject matter of this Agreement (together with Licensee, the "**Licensee Representatives**") that in connection with the performance of its obligations hereunder:

(i) The Licensee Representatives shall not directly or indirectly pay, offer or promise to pay or authorize the payment of any money or give, offer or promise

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to give or authorize the giving of anything else of value, to: (a) any Government Official in order to influence official action; (b) any Person (whether or not a Government Official) (I) to influence such Person to act in breach of a duty of good faith, impartiality or trust ("acting improperly"), (II) to reward such Person for acting improperly or (III) where such Person would be acting improperly by receiving the money or other thing of value; (c) any Person (whether or not a Government Official) while knowing or having reason to know that all or any portion of the money or other thing of value will be paid, offered, promised or given to or will otherwise benefit, a Government Official in order to influence official action for or against either Party in connection with the matters that are the subject of this Agreement; or (d) any Person (whether or not a Government Official) to reward that Person for acting improperly or to induce that Person to act improperly.

(ii) The Licensee Representatives shall not, directly or indirectly, solicit, receive or agree to accept any payment of money or anything else of value in violation of the Anti-Corruption Laws.

8.6.2. The Licensee Representatives shall comply with the Anti-Corruption Laws and shall not take any action that will, or would reasonably be expected to, cause MedImmune or its Affiliates to be in violation of any such laws.

8.6.3. Licensee shall promptly provide MedImmune with written notice of the following events: (i) upon becoming aware of any breach or violation by Licensee or other Licensee Representative of any representation, warranty or undertaking set out in Sections 8.6.1 through 8.6.2 above; or (ii) upon receiving a formal notification that it is the target of a formal investigation by a governmental authority for a Material Anti-Corruption Law Violation or upon receipt of information from any of the Licensee Representatives connected with this Agreement that any of them is the target of a formal investigation by a governmental authority for a Material Anti-Corruption Law Violation.

8.6.4. If MedImmune becomes aware that Licensee (or any other Licensee Representative) is in actual and definite breach or violation of any representation, warranty or undertaking in Sections 8.6.1 through 8.6.2 or of the Anti-Corruption Laws, MedImmune shall have the right, in addition to any other rights or remedies under this Agreement or to which MedImmune may be entitled in law or equity, to (i) take such steps, including by requiring Licensee to agree to such additional measures, representations, warranties, undertakings and other provisions, in each case, as MedImmune believes in good faith are reasonably necessary in order to avoid a potential violation or continuing violation by MedImmune or any of its Affiliates of the Anti-Corruption Laws ("**Provisions**") and (ii) terminate this Agreement terminate this Agreement for material breach in accordance with 10.2.2. in the event that:

(i) Licensee refuses to agree to all of the Provisions required by MedImmune pursuant to this clause; *provided* that MedImmune has (I) provided

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Licensee an explanation in reasonable detail as to why MedImmune considers such provisions necessary, (II) given Licensee a reasonable opportunity to review and comment on the proposed Provisions and to provide its view as to the necessity or usefulness of these to address the event concerned, and (III) considered such comments in good faith; or

(ii) MedImmune reasonably concludes that there is no Provision available that would enable MedImmune or its Affiliates to avoid a potential violation or continuing violation of applicable Anti-Corruption Laws.

8.6.5. Any termination of this Agreement pursuant to Section 8.6 shall be treated as a termination by MedImmune for Licensee's breach and the consequences of termination set out in Section 10.2.2 shall apply

8.6.6. Licensee shall be responsible for any breach of any representation, warranty or undertaking in this Section 8.6 or of the Anti-Corruption Laws by any Licensee Representative.

8.6.7. MedImmune may disclose the terms of this Agreement or any action taken under this Section 8.6 to prevent a potential violation or continuing violation of applicable Anti-Corruption Laws, including the identity of Licensee or a Licensee Representative and the payment terms, to any governmental authority if MedImmune determines, upon advice of counsel, that such disclosure is necessary.

ARTICLE 9
INDEMNITY

9.1. Indemnification by Licensee. Licensee shall indemnity MedImmune, its Affiliates, its or their (sub)licensees and its and their respective directors, officers, employees and agents and defend and save each of them harmless, from and against any and all losses, damages, liabilities, costs and expenses (including reasonable attorneys' fees and expenses), (collectively, "**Losses**") in connection with any and all suits, investigations, claims or demands of Third Parties (collectively, "**Third Party Claims**") arising from or occurring as a result of: (i) the breach by Licensee of this Agreement; (ii) the gross negligence or willful misconduct on the part of Licensee or its Affiliates or its or their Sublicensees or its or their distributors or contractors or its or their respective directors, officers, employees or agents in performing its or their obligations under this Agreement; or (iii) the Exploitation by Licensee or any of its Affiliates or its or their Sublicensees or its or their distributors or contractors of any Licensed Product or the Licensed Compound in or for the Territory, except, in each case ((i), (ii) and (iii)), for those Losses for which MedImmune has an obligation to indemnify Licensee pursuant to Section 9.2 hereof, as to which Losses each Party shall indemnify the other to the extent of their respective liability. hereof, as to which Losses each Party shall indemnify the other to the extent of their respective liability.

In calculating Losses, the legal duty to mitigate on the part of the Party suffering such Loss is taken into account.

9.2. Indemnification by MedImmune. MedImmune shall indemnify Licensee, its Affiliates and their respective directors, officers, employees and agents and defend and save each of them harmless, from and against any and all Losses in connection with any and all Third Party Claims arising from or occurring as a result of: (i) the breach by MedImmune of this Agreement (ii) the gross negligence or willful misconduct on the part of MedImmune or its Affiliates or its or their respective directors, officers, employees or agents in performing its obligations under this Agreement; (iii) the Exploitation of the Licensed Compound by or on behalf of MedImmune or its Affiliates prior to the Effective Date; and (iv) the Exploitation of any Licensed Compound or Licensed Product by or on behalf of MedImmune or its Affiliates following termination of this Agreement and/or any rights granted pursuant to Section 10.4.2; except, in each case ((i) — (iv)), for those Losses for which Licensee has an obligation to indemnify MedImmune pursuant to Section 9.1 hereof, as to which Losses each Party shall indemnify the other to the extent of their respective liability for the Losses.

9.3. Indemnification Procedures.

9.3.1. Notice of Claim. All indemnification claims in respect of a Party, its Affiliates or its or their (sub)licensees or their respective directors, officers, employees and agents shall be made solely by such Party to this Agreement (the "**Indemnified Party**"). The Indemnified Party shall give the indemnifying Party prompt written notice (an "**Indemnification Claim Notice**") of any Losses or discovery of fact upon which such indemnified Party intends to base a request for indemnification under this Article 9, but in no event shall the indemnifying Party be liable for any Losses that result from any delay in providing such notice. Each Indemnification Claim Notice must contain a description of the claim and the nature and amount of such Loss (to the extent that the nature and amount of such Loss is known at such time). The Indemnified Party shall furnish promptly to the indemnifying Party copies of all papers and official documents received in respect of any Losses and Third Party Claims.

9.3.2. Control of Defense. The indemnifying Party shall have the right to assume the defense of any Third Party Claim by giving written notice to the Indemnified Party within [***] days after the indemnifying Party's receipt of an Indemnification Claim Notice. The assumption of the defense of a Third Party Claim by the indemnifying Party shall not be construed as an acknowledgment that the indemnifying Party is liable to indemnify the Indemnified Party in respect of the Third Party Claim, nor shall it constitute a waiver by the indemnifying Party of any defenses it may assert against the Indemnified Party's claim for indemnification. Upon assuming the defense of a Third Party Claim, the indemnifying Party may appoint as lead counsel in the defense of the Third Party Claim any

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legal counsel selected by the indemnifying Party; *provided* that it obtains the prior written consent of the Indemnified Party (which consent shall not be unreasonably withheld, conditioned or delayed). In the event the indemnifying Party assumes the defense of a Third Party Claim, the Indemnified Party shall immediately deliver to the indemnifying Party all original notices and documents (including court papers) received by the Indemnified Party in connection with the Third Party Claim. Should the indemnifying Party assume the defense of a Third Party Claim, except as provided in Section 9.3.3, the indemnifying Party shall not be liable to the Indemnified Party for any legal expenses subsequently

incurred by such Indemnified Party in connection with the analysis, defense or settlement of the Third Party Claim unless specifically requested in writing by the indemnifying Party. In the event that it is ultimately determined that the indemnifying Party is not obligated to indemnify, defend or hold harmless the Indemnified Party from and against the Third Party Claim, the Indemnified Party shall reimburse the indemnifying Party for any and all reasonable and verifiable costs and expenses (including attorneys' fees and costs of suit) and any Losses incurred by the indemnifying Party in accordance with this Section 9 in its defense of the Third Party Claim.

9.3.3. Right to Participate in Defense. Any Indemnified Party shall be entitled to participate in the defense of such Third Party Claim and to employ counsel of its choice for such purpose; *provided*, *however*, that such employment shall be at the Indemnified Party's sole cost and expense unless (i) the employment thereof has been specifically authorized in writing by the indemnifying Party in writing (in which case, the defense shall be controlled as provided in Section 9.3.2), (ii) the indemnifying Party has failed to assume the defense and employ counsel in accordance with Section 9.3.2 (in which case the Indemnified Party shall control the defense) or (iii) the interests of the indemnitee and the indemnifying Party with respect to such Third Party Claim are sufficiently adverse to prohibit the representation by the same counsel of both Parties under Applicable Law, ethical rules or equitable principles (in which case, the Indemnified Party shall control its defense).

9.3.4. Settlement. With respect to any Losses relating solely to the payment of money damages in connection with a Third Party Claim and that shall not result in the applicable indemnitee(s) becoming subject to injunctive or other relief or otherwise adversely affecting the business of the Indemnified Party in any manner and as to which the indemnifying Party shall have acknowledged in writing the obligation to indemnify the applicable indemnitee hereunder, [***]. With respect to all other Losses in connection with Third Party Claims, where the indemnifying Party has assumed the defense of the Third Party Claim in accordance with Section 9.3.2, [***]. If the indemnifying Party does not assume and conduct the defense of a Third Party Claim as provided above, the Indemnified Party may defend against such Third Party Claim; *provided* that [***].

9.3.5. Cooperation. Regardless of whether the indemnifying Party chooses to defend or prosecute any Third Party Claim, the Indemnified Party shall and shall cause each indemnitee to, cooperate in the defense or prosecution thereof and shall furnish such records, information and testimony, provide such witnesses and attend such conferences, discovery proceedings, hearings, trials and appeals as may be reasonably requested in connection therewith. Such cooperation shall include access during normal business hours afforded to the indemnifying Party to and reasonable retention by the Indemnified Party of, records and information that are reasonably relevant to such Third Party Claim and making Indemnified Parties and other employees and agents available on a mutually convenient basis to provide additional information and explanation of any material provided hereunder and the indemnifying Party shall reimburse the Indemnified Party for all its, its Affiliates' and its and their (sub)licensees' or their respective directors', officers', employees' and agents', as applicable, reasonable and verifiable out-of-pocket expenses in connection therewith.

9.3.6. Expenses. Except as provided above, the costs and expenses, including fees and disbursements of counsel, incurred by the Indemnified Party and its Affiliates and its and their (sub)licensees and their respective directors, officers, employees and agents, as applicable, in connection with any claim shall be reimbursed on a Calendar Quarter basis by the indemnifying Party, without prejudice to the indemnifying Party's right to contest the Indemnified Party's right to indemnification and subject to refund in the event the indemnifying Party is ultimately held not to be obligated to indemnify the Indemnified Party.

9.4. Special, Indirect and Other Losses. EXCEPT (i) IN THE EVENT THE WILLFUL MISCONDUCT OR FRAUD OF A PARTY OR OF A PARTY'S BREACH OF ITS OBLIGATIONS UNDER ARTICLE 7, (ii) TO THE EXTENT ANY SUCH DAMAGES ARE REQUIRED TO BE PAID TO A THIRD PARTY AS PART OF A CLAIM FOR WHICH A PARTY PROVIDES INDEMNIFICATION UNDER THIS ARTICLE 9, NEITHER PARTY NOR ANY OF ITS AFFILIATES OR (SUB)LICENSEES SHALL BE LIABLE IN CONTRACT, TORT, NEGLIGENCE, BREACH OF STATUTORY DUTY OR OTHERWISE FOR ANY SPECIAL OR PUNITIVE DAMAGES OR FOR LOSS OF PROFITS SUFFERED BY THE OTHER PARTY. SHALL BE LIABLE IN CONTRACT, TORT, NEGLIGENCE, BREACH OF STATUTORY DUTY OR OTHERWISE FOR ANY SPECIAL OR PUNITIVE DAMAGES OR FOR LOSS OF PROFITS SUFFERED BY THE OTHER PARTY.

9.5. Insurance. Licensee shall have and maintain such types and amounts of insurance covering its Exploitation of the Licensed Products as is (i) [***] and (ii) otherwise required by Applicable Law.

ARTICLE 10
TERM AND TERMINATION

10.1. Term and Expiration. This Agreement commences on the Effective Date and, unless earlier terminated in accordance with this Article 10, continues in force and effect until the date of expiration of the Royalty Term in the last country for the last Indication in the Territory, (such period, the "**Term**"). Following the expiration of the Royalty Term for a Licensed Product in a country, the grants in Section 2.1 shall become fully-paid, royalty-free, and irrevocable for such Licensed Product in such country. Upon the expiration of the Term, the grants in Section 2.1 shall become non-exclusive, fully-paid royalty-free, and irrevocable.

10.2. Termination.

10.2.1. By Licensee for Convenience. Licensee may terminate this Agreement in its entirety for convenience upon providing ninety (90) days prior written notice to MedImmune.

10.2.2. For Material Breach. In the event that either Party (the "**Breaching Party**") is in material breach in the performance of any of its obligations under this Agreement, in addition to any other right and remedy the other Party (the "**Non-Breaching Party**") may have, the Non-Breaching Party may terminate this Agreement in its entirety, by providing ninety (90) days (the "**Notice Period**") prior written notice (the "**Termination Notice**") to the Breaching Party and specifying the breach and its claim of right to terminate; *provided* that (i) the termination shall not become effective at the end of the Notice Period if the Breaching Party cures the breach specified in the Termination Notice during the Notice Period (or, if such default cannot be cured within the Notice Period, if the Breaching Party commences actions to cure such breach within the Notice Period and thereafter diligently continues such actions)and (ii) with respect to an uncured material breach consisting of Licensee's diligence obligations under Section 4.1.1 or Section 4.3.1, as applicable,. For the purposes of termination "**material breach"** means a breach of obligations under this Agreement where such breach has a significant adverse effect on the other Party's rights and obligations under this Agreement, including but not limited to, uncured non-payment of milestones and royalties and acts or omissions that result the inability of a Party to continue with the Development Plan and/or the Commercialization Pan for the Licensed Product. If there is a *bona fide* dispute between the Parties as to whether any such material breach has occurred and/or as to the nature of a breach being a material breach, the Parties will resolve such dispute in good faith in accordance with Section 11.5 (Dispute Resolution). During such dispute resolution procedure to determine whether a material breach has occurred, neither Party may terminate the Agreement.

10.2.3. Termination by MedImmune for Patent Challenge. In the event that Licensee or any of its Affiliates or Sublicensees, anywhere in the Territory, institutes,

prosecutes or otherwise participates in (or in any way aids any Third Party in instituting, prosecuting or participating in), at law or in equity or before any administrative or regulatory body, including the U.S. Patent and Trademark Office or its foreign counterparts, any claim, demand, action or cause of action for declaratory relief, damages or any other remedy or for an enjoinment, injunction or any other equitable remedy, including any interference, re-examination, opposition or any similar proceeding, alleging that any claim in a Licensed Patent is invalid, unenforceable or otherwise not patentable or would not be infringed by Licensee's activities absent the rights and licenses granted hereunder, MedImmune shall have the right to immediately terminate this Agreement in its entirety, including the rights of any Sublicensees, upon written notice to Licensee; *provided*, *however*, that MedImmune will not have the right to terminate this Agreement under this Section 10.2.3 for any such challenge by any Sublicensee if (i) Licensee terminates the Sublicense within [***]days of MedImmune's notice to Licensee, or (ii) such challenge is dismissed within [***] days of MedImmune's notice to Licensee under this Section 10.2.3 and not thereafter continued.

10.2.4. Termination for Insolvency. In the event that either Party (i) files for protection under bankruptcy or insolvency laws, (ii) makes an assignment for the benefit of creditors, (iii) appoints or suffers appointment of a receiver or trustee over substantially all of its property that is not discharged within [***] days after such filing, (iv) proposes a written agreement of composition or extension of its debts, (v) proposes or is a party to any dissolution or liquidation, (vi) files a petition under any bankruptcy or insolvency act or has any such petition filed against that is not discharged within [***] days of the filing thereof or (vii) admits in writing its inability generally to meet its obligations as they fall due in the general course, then the other Party may terminate this Agreement in its entirety effective immediately upon written notice to such Party.

10.3. Rights in Bankruptcy. All rights and licenses granted under or pursuant to this Agreement by Licensee or MedImmune are and shall otherwise be deemed to be, for purposes of Section 365(n) of the U.S. Bankruptcy Code or any analogous provisions in any other country or jurisdiction, licenses of right to "intellectual property" as defined under Section 101 of the U.S. Bankruptcy Code. The Parties agree that the Parties, as licensees of such rights under this Agreement, shall retain and may fully exercise all of their rights and elections under the U.S. Bankruptcy Code or any analogous provisions in any other country or jurisdiction. The Parties further agree that, in the event of the commencement of a bankruptcy proceeding by or against either Party under the U.S. Bankruptcy Code or any analogous provisions in any other country or jurisdiction, the Party hereto that is not a Party to such proceeding shall be entitled to a complete duplicate of (or complete access to, as appropriate) any such intellectual property and all embodiments of such intellectual property, which, if not already in the non-subject Party's possession, shall be promptly delivered to it (i) upon any such commencement of a bankruptcy proceeding upon the non-subject Party's written request therefor, unless the Party subject to such proceeding elects to continue to perform all of its obligations under this Agreement, or (ii) if

not delivered under clause (i) above, following the rejection of this Agreement by or on behalf of the Party subject to such proceeding upon written request therefor by the non-subject Party.

10.4. Consequences of Termination.

10.4.1. Termination of the Agreement. In the event of a termination of this Agreement in its entirety:

(i) **By Licensee for Convenience**. Sections 10.4.2 (i) through (viii) will apply.

(ii) **By Licensee for Material Breach by MedImmune or MedImmune's insolvency under 10.2.4**. In the event of a Material breach of this Agreement by MedImmune, or MedImmune becomes insolvent, Licensee shall have the option to (a) terminate the Agreement and return the Licensed Technology and Licensed Products to MedImmune in accordance with Sections 10.4.2 (i) through (vii); or (b) retain Licensed Technology and Licensed Product and continue to Exploit the Licensed Products, whereby all payment obligations from Licensee to MedImmune shall be reduced by [***]. Such reduction in milestones and royalties will be Licensee's sole remedy. [***].

(iii) **By MedImmune for Material Breach by Licensee or Licensee's insolvency**. Sections 10.4.2 (i) through (viii) will apply.

10.4.2. Applicable Termination Provisions. The following shall apply to the termination events as described above:

(i) all rights and licenses granted by MedImmune hereunder shall immediately terminate;

(ii) Licensee shall and hereby does, and shall cause its Affiliates and its and their Sublicensees to, effective as of the effective date of termination assign to MedImmune all of its right, title and interest in and to (a) each Product Trademark and (b) all Regulatory Documentation (including any Regulatory Approvals) applicable to any Licensed Compound or Licensed Products or any Improvement thereto then owned or Controlled by Licensee or any of its Affiliates; *provided* that if any such Regulatory Documentation or Regulatory Approval is not immediately transferable in a country, Licensee shall provide MedImmune with all benefit of such Regulatory Documentation or Regulatory Approval, as applicable, and such assistance and cooperation as necessary or reasonably requested by MedImmune to timely transfer such Regulatory Documentation or Regulatory Approval, as applicable, to MedImmune or its designee [***];

(iii) all Confidential Information of Licensee directly and solely relating to the Licensed Compound or any Licensed Product shall become Confidential Information of MedImmune;

(iv) Licensee shall and hereby does, and shall cause its Affiliates and its and their Sublicensees to, effective as of the effective date of termination, grant MedImmune an exclusive, license and right of reference, with the right to grant multiple tiers of sublicenses and further rights of reference, in and to the Licensee Intellectual Property. that are not assigned to MedImmune pursuant to clause (ii) above, to Exploit in the Territory any Licensed Compound or Licensed Product;

(v) unless expressly prohibited by any Regulatory Authority, at MedImmune's written request, Licensee shall and hereby does, and shall cause its Affiliates and its and their Sublicensees to, (a) transfer control to MedImmune [***] clinical studies involving Licensed Products or any Improvements thereto being conducted by or on behalf of Licensee, an Affiliate or a Sublicensee as of the effective date of termination and (b) continue to conduct such clinical studies, [***], for up to [***] months to enable such transfer to be completed without interruption of any such clinical study; *provided* that (x) MedImmune shall not have any obligation to continue any clinical study unless required by Applicable Law and (y) with respect to each clinical study for which such transfer is expressly prohibited by the applicable Regulatory Authority, if any, Licensee shall continue to conduct such clinical study [***];

(vi) at MedImmune's written request, Licensee shall, and cause its Affiliates and its and their Sublicensees to, assign to MedImmune all Licensed Product Agreements, unless, with respect to any such Licensed Product Agreement, such Licensed Product Agreement expressly prohibits such assignment, in which case Licensee (or such Affiliate or Sublicensee, as applicable) shall cooperate with MedImmune in all reasonable respects to secure the consent of the applicable Third Party to such assignment and if any such consent cannot be obtained with respect to a Licensed Product Agreement, Licensee shall, and cause its Affiliates and its and their Sublicensees to, obtain for MedImmune substantially all of the practical benefit and burden under such Licensed Product Agreement, including by (a) entering into appropriate and reasonable alternative arrangements on terms agreeable to MedImmune and (b) subject to the consent and control of MedImmune, enforcing, at MedImmune's cost and expense and for the account of MedImmune, any and all rights of Licensee (or such Affiliate or Sublicensee, as applicable) against the other party thereto arising out of the breach or cancellation thereof by such other party or otherwise;

(vii) at MedImmune' written request, Licensee shall supply to MedImmune such quantities of the Licensed Compound and Licensed Products as MedImmune indicates in written forecasts and orders [***] pursuant to a supply agreement

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to be negotiated in good faith between the Parties to Manufacture such Licensed Compound and Licensed Products until [***] (a) [***], (b) [***] (c) [***]; provided that MedImmune promptly commence, and take steps to completed a process to establish such alternative source as soon as reasonably possible; and

(viii) In the event the Agreement is terminated by Licensee for convenience or by Medimmune for Licensee's Material Breach, or by MedImmune under Section 10.2.3, Licensee will grant to MedImmune a non-exclusive, royalty free, subslicensable right and license to the Licensee Intellectual Property necessary for and actually used in connection with the Licensed Product to enable MedImmune to further Exploit the Licensed Product. Should MedImmune desire an exclusive license to the Licensee Intellectual Property in order to Exploit the Licensed Product, the Parties will negotiate in good faith the terms of a royalty-bearing exclusive license to use such Licensee Intellectual Property. In addition, should such termination take place after the Licensee has received Regulatory Approval of the Product in any Indication, the Parties shall negotiate in good faith a royalty on Net Sales of Licensed Product to Licensee [***]. If the Parties are unable to reach agreement on such terms, then either Party may elect to have such matter resolved subject to the dispute resolution provisions set forth in Section 11.5.

(ix) In the event Licensee terminates this Agreement for Material Breach by MedImmune, and does not wish to take its option in Section 10.4.1(ii) to continue Exploiting the Licensed Product, and if MedImmune wishes to Exploit the Licensed Product, Licensee will grant to MedImmune a royalty-bearing, non-exclusive or exclusive license as the case may be to the Licensee Intellectual Property and the Parties will [***] in good faith [***]. In addition, should such termination take place after the Licensee has received Regulatory Approval of the Product in an Indication, the Parties shall [***] to Licensee [***]. If the Parties are unable to reach agreement on such terms, then either Party may elect to have such matter resolved subject to the dispute resolution provisions set forth in Section 11.5.

10.5. Accrued Rights; Surviving Obligations. Termination or expiration of this Agreement for any reason shall be without prejudice to any rights that shall have accrued to the benefit of a Party prior to such termination or expiration. Such termination or expiration shall not relieve a Party from obligations that are expressly indicated to survive the termination or expiration of this Agreement. Without limiting the foregoing, Articles 1, 7, 9, 10 and Sections 11.2, and 11.5 through 11.11 of this Agreement shall survive the termination or expiration of this Agreement for any reason.

ARTICLE 11
MISCELLANEOUS

11.1. Force Majeure. Neither Party shall be held liable or responsible to the other Party or be deemed to have defaulted under or breached this Agreement for failure or delay

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in fulfilling or performing any term of this Agreement (other than an obligation to make payments) when such failure or delay is caused by or results from events beyond the reasonable control of the non-performing Party, including fires, floods, earthquakes, hurricanes, embargoes, shortages, epidemics, quarantines, war, acts of war (whether war be declared or not), terrorist acts, insurrections, riots, civil commotion, strikes, lockouts or other labor disturbances (whether involving the workforce of the non-performing Party or of any other Person), acts of God or acts, omissions or delays in acting by any governmental authority (except to the extent such delay results from the breach by the non-performing Party or any of its Affiliates of any term or condition of this Agreement). The non-performing Party shall notify the other Party of such force majeure within [***] days after such occurrence by giving written notice to the other Party stating the nature of the event, its anticipated duration and any action being taken to avoid or minimize its effect. The suspension of performance shall be of no greater scope and no longer duration than is necessary and the non-performing Party shall use commercially reasonable efforts to remedy its inability to perform.

11.2. Export Control. This Agreement is made subject to any restrictions concerning the export of products or technical information from the United States or other countries that may be imposed on the Parties from time to time. Each Party agrees that it will not export, directly or indirectly, any technical information acquired from the other Party under this Agreement or any products using such technical information to a location or in a manner that at the time of export requires an export license or other governmental approval, without first obtaining the written consent to do so from the appropriate agency or other governmental entity in accordance with Applicable Law. This Agreement is made subject to any restrictions concerning the export of products or technical information from the United States or other countries that may be imposed on the Parties from time to time. Each Party agrees that it will not export, directly or indirectly, any technical information acquired from the other Party under this Agreement or any products using such technical information to a location or in a manner that at the time of export requires an export license or other governmental approval, without first obtaining the written consent to do so from the appropriate agency or other governmental entity in accordance with Applicable Law.

11.3. Assignment.

11.3.1. Neither Party may assign its rights whether by operation of law or otherwise, in whole or in part without the prior written consent of the other Party, which consent shall not be unreasonably withheld, conditioned or delayed, except that the Party's shall have the right, without such consent, (i) to perform any or all of its obligations and exercise any or all of its rights under this Agreement through any of its Affiliates or its or their (sub)licensees, and

(ii) assign any or all of its rights and delegate any or all of its obligations hereunder to any of its Affiliates or its or their (sub)licensees, or to any successor in interest (whether by merger, acquisition, asset purchase or otherwise) to all or

substantially all of the business to which this Agreement relates; *provided* that MedImmune shall provide written notice to Licensee within [***] days after such assignment. Any permitted successor of a Party or any permitted assignee of all of a Party's rights under this Agreement that has also assumed all of such Party's obligations hereunder in writing shall, upon any such succession or assignment and assumption, be deemed to be a party to this Agreement as though named herein in substitution for the assigning Party, whereupon the assigning Party shall cease to be a party to this Agreement and shall cease to have any rights or obligations under this Agreement. All validly assigned rights of a Party shall inure to the benefit of and be enforceable by, and all validly delegated obligations of such Party shall be binding on and be enforceable against, the permitted successors and assigns of such Party; *provided* that such Party, if it survives, shall remain jointly and severally liable for the performance of such delegated obligations under this Agreement. Any attempted assignment or delegation in violation of this Section 11.3.1 shall be void and of no effect. Notwithstanding the foregoing, in the event a Party assigns its rights and obligations under this Agreement or otherwise makes Payments from a jurisdiction in which such Party is organized (each an "Assignment"), and [***].

11.3.2. The rights to Information, materials and Intellectual Property: (i) controlled by a Third Party permitted assignee of a Party that immediately prior to such assignment (other than as a result of a license or other grant of rights, covenant or assignment by such Party or its Affiliates to, or for the benefit of, such Third Party); or (ii) controlled by an Affiliate of a Party that becomes an Affiliate through any Change of Control of such Party that were controlled by such Affiliate (and not such Party) immediately prior to such Change of Control (other than as a result of a license or other grant of rights, covenant or assignment by such Party or its other Affiliates to, or for the benefit of, such Affiliate), in each case ((i) and (ii)), [***].

11.4. Severability. If any provision of this Agreement is held to be illegal, invalid or unenforceable under any present or future law and if the rights or obligations of either Party under this Agreement will not be materially and adversely affected thereby, (i) such provision shall be fully severable, (ii) this Agreement shall be construed and enforced as if such illegal, invalid or unenforceable provision had never comprised a part hereof, (iii) the remaining provisions of this Agreement shall remain in full force and effect and shall not be affected by the illegal, invalid or unenforceable provision or by its severance herefrom, and (iv) in lieu of such illegal, invalid or unenforceable provision, there shall be added automatically as a part of this Agreement a legal, valid and enforceable provision as similar in terms to such illegal, invalid or unenforceable provision as may be possible and reasonably acceptable to the Parties. To the fullest extent permitted by Applicable Law, each Party hereby waives any provision of law that would render any provision hereof illegal, invalid or unenforceable in any respect. unenforceable in any respect.

11.5. Dispute Resolution.

11.5.1. If a dispute arises between the Parties in connection with or relating to this Agreement or any document or instrument delivered in connection herewith (collectively, (i) and (ii), a "**Dispute**"), then either Party shall have the right to refer such Dispute to the Senior Officers for attempted resolution by good faith negotiations during a period of [***] Business Days. Any final decision mutually agreed to by the Executive Officers shall be conclusive and binding on the Parties.

11.5.2. If such Executive Officers are unable to resolve any such Dispute within such [***] Business Day period, either Party shall be free to institute binding arbitration in accordance with this Section 11.5.2 upon written notice to the other Party (an "**Arbitration Notice**") and seek such remedies as may be available. Upon receipt of an Arbitration Notice by a Party, the applicable Dispute shall be resolved by final and binding arbitration before a panel of three (3) experts with relevant industry experience (the "**Arbitrators**"). The arbitration shall be administered by the American Arbitration Association in accordance with the then current Commercial Rules of the American Arbitration Association including the Procedures for Large, Complex Commercial Disputes (including the Optional Rules for Emergency Measures of Protection) ("**AAA Rules**"), except that any such arbitration must be conducted in accordance with the remainder of this Section 11.5. Each of Licensee and MedImmune shall promptly select one (1) Arbitrator, which selections shall in no event be made later than [***] days after the notice of initiation of arbitration. The third Arbitrator shall be chosen promptly by mutual agreement of the Arbitrator chosen by Licensee and the Arbitrator chosen by MedImmune, but in no event later than [***] days after the date that the last of such Arbitrators was appointed. The third appointed arbitrator shall serve as the chairman arbitrator and the chairman shall be a lawyer admitted to practice in New York, USA for at least fifteen (15) years, and who is experienced with disputes in Licensing transactions. The Arbitrators shall determine what discovery will be permitted, consistent with the goal of reasonably controlling the cost and time that the Parties must expend for discovery; *provided* that the Arbitrators shall permit such discovery as they deem necessary to permit an equitable resolution of the dispute. The place of arbitration shall be New York, USA at a suitable venue to be agreed by the parties and arbitrators within [***] Business Days of the appointment of the chairman arbitrator. The proceedings shall be conducted in the English language. The Parties shall use reasonable efforts to expedite the arbitration if requested by either Party. The Arbitrators shall, within [***] days after the conclusion of the arbitration hearing, issue a written award and statement of decision describing the essential findings and conclusions on which the award is based, including the calculation of any damages awarded. The decision or award rendered by the Arbitrators shall be final and non-appealable, and judgment may be entered upon it in accordance with Applicable Law in England and Wales or any other court of competent jurisdiction. The Arbitrators shall be authorized to award compensatory damages, but shall not be authorized to reform, modify or materially change this Agreement or any other agreements contemplated hereunder.

11.5.3. Each Party shall bear its own counsel fees, costs, and disbursements arising out of the dispute resolution procedures described in this Section 11.5, and shall pay an equal share of the fees and costs of the Expert or Arbitrators, as applicable, and all other general fees related to any arbitration described in Section 11.5.2; *provided, however*, the Arbitrators shall be authorized to determine whether a Party is the prevailing Party, and if so, to award to that prevailing Party reimbursement for its reasonable counsel fees, costs and disbursements (including expert witness fees and expenses, photocopy charges, or travel expenses), or the fees and costs of the Arbitrators. Unless the Parties otherwise agree in writing, during the period of time that any arbitration proceeding described in Section 11.5.2 is pending under this Agreement, the Parties shall continue to comply with all those terms and provisions of this Agreement that are not the subject of such pending arbitration proceeding. Nothing contained in this Agreement shall deny any Party the right to seek injunctive or other equitable relief from a court of competent jurisdiction in the context of a bona fide emergency or prospective irreparable harm, and such an action may be filed and maintained notwithstanding any ongoing arbitration proceeding. All arbitration proceedings and decisions of the Expert or Arbitrator, as applicable, under this 11.5.3 shall be deemed Confidential Information of both Parties under Article 7.

11.6. Governing Law. This Agreement shall be governed by and construed in accordance with the laws of [***] excluding any conflicts or choice of law rule or principle that might otherwise refer construction or interpretation of this Agreement to the substantive law of another jurisdiction.

11.7. Notices.

11.7.1. Notice Requirements. Any notice, request, demand, waiver, consent, approval or other communication permitted or required under this Agreement shall be in writing, shall refer specifically to this Agreement and shall be deemed given only if delivered by hand or sent by facsimile transmission (with transmission confirmed) or by internationally recognized overnight delivery service that maintains records of delivery, addressed to the Parties at their respective addresses specified in Section 11.7.2 or to such other address as the Party to whom notice is to be given may have provided to the other Party in accordance with this Section 11.7.1. Such Notice shall be deemed to have been given as of the date delivered by hand or transmitted by facsimile (with transmission confirmed) or on the second Business Day (at the place of delivery) after deposit with an internationally recognized overnight delivery service. Any notice delivered by facsimile shall be confirmed by a hard copy delivered as soon as practicable thereafter. This Section 11.7.1 is not intended to govern the day-to-day business communications necessary between the Parties in performing their obligations under the terms of this Agreement.

11.7.2. Address for Notice.

If to Licensee:
Kiniksa Pharmaceuticals, Ltd.
Clarendon House
2 Church Street
Hamilton HM 11
Bermuda
Attention: Chief Legal Officer

With copies to:
Kiniksa Pharmaceuticals Corp.
15 Walnut Street
Wellesley, MA 02481
Attention: Legal Department

If to MedImmune, to:

Alliance Management
MedImmune
Milstein Building, Granta Park
Cambridge, CB21 6GH, UK

With copies to:

Deputy General Counsel, Corporate
MedImmune
Milstein Building, Granta Park
Cambridge, CB21 6GH, UK

11.8. Entire Agreement; Amendments. This Agreement, together with the Schedules attached hereto, sets forth and constitutes the entire agreement and understanding between the Parties with respect to the subject matter hereof and all prior agreements, understandings, promises and representations, whether written or oral, with respect thereto are superseded hereby. Each Party confirms that it is not relying on any representations or warranties of the other Party except as specifically set out in this Agreement. No amendment, modification, release or discharge shall be binding on the Parties unless in writing and duly executed by authorized representatives of both Parties. In the event of any inconsistencies between this Agreement and any schedules or other attachments hereto, the terms of this Agreement shall control.

11.9. English Language. This Agreement shall be written and executed in and all other communications under or in connection with this Agreement shall be in, the English language. Any translation into any other language shall not be an official version thereof

and in the event of any conflict in interpretation between the English version and such translation, the English version shall control. in the event of any conflict in interpretation between the English version and such translation, the English version shall control.

11.10. Equitable Relief. In the event of a breach or threatened breach of any provision of Article 7, the non-breaching Party shall be authorized and entitled to seek from any court of competent jurisdiction injunctive relief, whether preliminary or permanent, which rights shall be cumulative and in addition to any other rights or remedies to which such non-breaching Party may be entitled in law or equity. Both Parties agree to waive any requirement that the other (i) post a bond or other security as a condition for obtaining any such relief and (ii) show irreparable harm, balancing of harms, consideration of the public interest or inadequacy of monetary damages as a remedy. Nothing in this Section 11.10 should be construed, to limit either Party's right to equitable relief or any other remedy for a breach of any other provision of this Agreement.

11.11. Waiver and Non-Exclusion of Remedies. Any term or condition of this Agreement may be waived in writing at any time by the Party that is entitled to the benefit thereof, but no such waiver shall be effective unless set out in a written instrument duly executed by or on behalf of the Party waiving such term or condition. The waiver by either Party hereto of any right hereunder or of the failure to perform or of a breach by the other Party shall not be deemed a waiver of any other right hereunder or of any other breach or failure by such other Party whether of a similar nature or otherwise. The rights and remedies provided herein are cumulative and do not exclude any other right or remedy provided by Applicable Law or otherwise available except as expressly set out herein.

11.12. No Benefit to Third Parties. Except for any rights and immunities granted in this Agreement to any Affiliates, the Contracts (Rights of Third Parties) Act 1999 shall not apply to this Agreement. Except as expressly provided in Article 10, no Person who is not a party to this Agreement (including any employee, officer, agent, representative or subcontractor of either Party) shall have the right (whether under the Contracts (Rights of Third Parties) Act 1999 or otherwise) to enforce any provision of this Agreement which expressly or by implication confers a benefit on that Person without the express prior agreement in writing of the Parties, which agreement must refer to this Section 11.12.

11.13. Further Assurance. Each Party shall duly execute and deliver or cause to be duly executed and delivered, such further instruments and do and cause to be done such further acts and things, including the filing of such assignments, agreements, documents and instruments, as may be necessary or as the other Party may reasonably request in connection with this Agreement or to carry out more effectively the provisions and purposes hereof or to better assure and confirm unto such other Party its rights and remedies under this Agreement.

11.14. Relationship of the Parties. MedImmune, on the one hand and Licensee, on the other hand, shall be independent contractors and that the relationship between the two Parties shall not constitute a partnership, joint venture or agency. Neither MedImmune, on the one hand, nor Licensee, on the other hand, shall have the authority to make any statements, representations or commitments of any kind or to take any action, that will be binding on the other, without the prior written consent of the other Party to do so. All persons employed by a Party shall be employees of such Party and not of the other Party and all costs and obligations incurred by reason of any such employment shall be for the account and expense of such first Party.

11.15. Non-Solicitation of Employees. Commencing on the Effective Date and for a period of [***] thereafter, neither Party shall, directly or indirectly, actively recruit or solicit any then-current employee of the other Party with whom such Party has come into contact or interacted for the purposes of performing this Agreement, without the prior consent of the other Party. For purposes of this Section, "solicit" shall be deemed not to include: (i) circumstances where an employee of one Party or any of its Affiliates initially contacts the other Party or any of such Party's Affiliates seeking employment; or (ii) general solicitations of employment not specifically targeted at such employees.

11.16. References. Unless otherwise specified, (i) references in this Agreement to any Article, Section or Schedule shall mean references to such Article, Section or Schedule of this Agreement, (ii) references in any Section to any clause are references to such clause of such Section and (iii) references to any agreement, instrument or other document in this Agreement refer to such agreement, instrument or other document as originally executed or, if subsequently amended, replaced or supplemented from time to time, as so amended, replaced or supplemented and in effect at the relevant time of reference thereto.

11.17. Construction. Except where the context otherwise requires, wherever used, the singular shall include the plural, the plural the singular, the use of any gender shall be applicable to all genders and the word "or" is used in the inclusive sense (and/or). Whenever this Agreement refers to a number of days, unless otherwise specified, such number refers to calendar days. The captions of this Agreement are for convenience of reference only and in no way define, describe, extend or limit the scope or intent of this Agreement or the intent of any provision contained in this Agreement. The term "including," "include," or "includes" as used herein shall mean including, without limiting the generality of any description preceding such term. The language of this Agreement shall be deemed to be the language mutually chosen by the Parties and no rule of strict construction shall be applied against either Party hereto. limit the scope or intent of this Agreement or the intent of any provision contained in this Agreement. The term "including," "include," or "includes" as used herein shall mean including, without limiting the generality of any description preceding such term.

The language of this Agreement shall be deemed to be the language mutually chosen by the Parties and no rule of strict construction shall be applied against either Party hereto.

11.18. Counterparts. This Agreement may be executed in two (2) or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument. This Agreement may be executed by facsimile, PDF format via email or other electronically transmitted signatures and such signatures shall be deemed to bind each Party hereto as if they were original signatures electronically transmitted signatures and such signatures shall be deemed to bind each Party hereto as if they were original signatures.

[SIGNATURE PAGE FOLLOWS.**]**

THIS AGREEMENT IS EXECUTED by the authorized representatives of the Parties as of the date first written above.

MEDIMMUNE, LIMITED	**KINIKSA PHARMACEUTICALS, LTD.**
By: [***]	By: /s/ Thomas Beetham

Name:	[***]	Name:	Thomas Beetham
Title:	[***]	Title:	Executive Vice President

SCHEDULES

Schedule 1.39	**Exclusive Licensed Patents**
Schedule 1.56	**In-License Agreements**
Schedule 1.74	**Non-Exclusive Licensed Patents**
Schedule 1.104	**Transition Activities**
Schedule 3.1	**Purchased Assets**
Schedule 8.2.3	**Actions for Exclusive Licensed Patents as at the Effective Date**

Schedule 1.39

[***]

Schedule 1.56

[***]

Schedule 1.74

[***]

Schedule 1.104
Transition Activities

[***]

Schedule 3.1 Inventory

[***]

Schedule 8.2.3

[***]

KINIKSA PHARMACEUTICALS, LTD.
2018 EMPLOYEE SHARE PURCHASE PLAN

ARTICLE I.
PURPOSE

The purposes of this Kiniksa Pharmaceuticals, Ltd. 2018 Employee Share Purchase Plan (as it may be amended or restated from time to time, the "***Plan***") are to assist Eligible Employees of Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company (the "***Company***"), and its Designated Subsidiaries in acquiring an ownership interest in the Company pursuant to a plan which is intended to qualify as an "employee stock purchase plan" within the meaning of Section 423(b) of the Code, and to help Eligible Employees provide for their future security and to encourage them to remain in the employment of the Company and its Designated Subsidiaries.

ARTICLE II.
DEFINITIONS AND CONSTRUCTION

Wherever the following terms are used in the Plan they shall have the meanings specified below, unless the context clearly indicates otherwise. The singular pronoun shall include the plural where the context so indicates. Masculine, feminine and neuter pronouns are used interchangeably and each comprehends the others.

2.1 "***Administrator***" shall mean the entity that conducts the general administration of the Plan as provided in Article XI. The term "Administrator" shall refer to the Committee unless the Board has assumed the authority for administration of the Plan as provided in Article XI.

2.2 "***Applicable Law***" shall mean the requirements relating to the administration of equity incentive plans under U.S. federal and state securities, tax and other applicable laws, rules and regulations, the applicable rules of any stock exchange or quotation system on which the Common Shares are listed or quoted and the applicable laws and rules of any foreign country or other jurisdiction where rights under this Plan are granted, including without limitation, the laws of Bermuda.

2.3 "***Board***" shall mean the Board of Directors of the Company.

2.4 "***Change in Control***" shall mean (a) a sale of all or substantially all of the Company's assets, or (b) any merger, amalgamation, consolidation or other business combination transaction of the Company with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the voting shares of the Company outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into voting shares of the surviving entity) a majority of the total voting power represented by the voting shares of the Company (or the surviving entity) outstanding immediately after such transaction, or (c) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of the Company. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur: (A) on account of the acquisition of voting shares by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Company's voting shares in a transaction or series of related transactions that are primarily a private financing transaction for the Company or (B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or

persons acting as a group (the "***Subject Person***"), exceeds the designated percentage threshold of the outstanding voting shares as a result of a repurchase or other acquisition of voting shares by the Company reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operating of this sentence) as a result of the acquisition of voting shares by the Company, and after such share acquisition, the Subject Person becomes the owner of any additional voting shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

The Administrator shall have full and final authority, which shall be exercised in its discretion, to determine conclusively whether a Change in Control has occurred pursuant to the above definition, the date of the occurrence of such Change in Control and any incidental matters relating thereto.

2.5 "***Code***" shall mean the Internal Revenue Code of 1986, as amended, and the regulations issued thereunder.

2.6 "***Common Shares***" shall mean the Class A Common Shares of the Company of par value US$1.0001 each.

2.7 "***Company***" shall mean of Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company, or any successor.

2.8 "***Compensation***" of an Eligible Employee shall mean the gross base compensation received by such Eligible Employee as compensation for services to the Company or any Designated Subsidiary, including overtime payments and excluding sales commissions, incentive compensation, bonuses, expense reimbursements, fringe benefits and other special payments.

2.9 "***Designated Subsidiary***" shall mean any Subsidiary designated by the Administrator in accordance with Section 11.3(b).

2.10 "***Effective Date***" shall mean the day prior to the Public Trading Date.

2.11 "***Eligible Employee***" shall mean an Employee: (a) who does not, immediately after any rights under this Plan are granted, own (directly or through attribution) shares possessing 5% or more of the total combined voting power or value of all classes of Common Shares and other shares of the Company, a Parent or a Subsidiary (as determined under Section 423(b)(3) of the Code); (b) whose customary employment is for more than twenty hours per week; and (c) whose customary employment is for more than five months in any calendar year. For purposes of the foregoing, the rules of Section 424(d) of the Code with regard to the attribution of share ownership shall apply in determining the share ownership of an individual, and shares that an Employee may purchase under outstanding options shall be treated as shares owned by the Employee; provided, however, that the Administrator may provide in an Offering Document that an Employee shall not be eligible to participate in an Offering Period if: (i) such Employee is a highly compensated employee within the meaning of Section 423(b)(4)(D) of the Code; and/or (ii) such Employee has not met a service requirement designated by the Administrator pursuant to Section 423(b)(4)(A) of the Code (which service requirement may not exceed two years), and/or (iii) such Employee is a citizen or resident of a foreign jurisdiction and the grant of a right to purchase Common Shares under the Plan to such Employee would be prohibited under the laws of such foreign jurisdiction or the grant of a right to purchase Common Shares under the Plan to such Employee in compliance with the laws of such foreign jurisdiction would cause the Plan to violate the requirements of Section 423 of the Code, as determined by the Administrator in its sole discretion; provided, further, that any exclusion in clauses (i), (ii) or (iii) shall

be applied in an identical manner under each Offering Period to all Employees, in accordance with Treasury Regulation Section 1.423-2(e).

2.12 "***Employee***" shall mean any officer or other employee (as defined in accordance with Section 3401(c) of the Code) of the Company or any Designated Subsidiary. "Employee" shall not include any director of the Company or a Designated Subsidiary who does not render services to the Company or a Designated Subsidiary as an employee within the meaning of Section 3401(c) of the Code. For purposes of the Plan, the employment relationship shall be treated as continuing intact while the individual is on sick leave or other leave of absence approved by the Company or Designated Subsidiary and meeting the requirements of Treasury Regulation Section 1.421-1(h)(2). Where the period of leave exceeds three (3) months and the individual's right to reemployment is not guaranteed either by statute or by contract, the employment relationship shall be deemed to have terminated on the first day immediately following such three (3)-month period.

2.13 "***Enrollment Date***" shall mean the first Trading Day of each Offering Period.

2.14 "***Exchange Act***" shall mean the Securities Exchange Act of 1934, as amended.

2.15 "***Fair Market Value***" means, as of any date, the value of Common Shares determined as follows: (i) if the Common Shares are listed on any established stock exchange, a share's Fair Market Value will be the closing sales price for such Common Share as quoted on such exchange for such date, or if no sale occurred on such date, the last day preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; (ii) if the Common Shares are not traded on a stock exchange but is quoted on a national market or other quotation system, the closing sales price on such date, or if no sales occurred on such date, then on the last date preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; or (iii) without an established market for the Common Shares, the Administrator will determine the Fair Market Value in its discretion.

2.16 "***Offering Document***" shall have the meaning given to such term in Section 4.1.

2.17 "***Offering Period***" shall have the meaning given to such term in Section 4.1.

2.18 "***Parent***" shall mean any corporation, other than the Company, in an unbroken chain of corporations ending with the Company if, at the time of the determination, each of the corporations other than the Company owns shares possessing 50% or more of the total combined voting power of all classes of shares in one of the other corporations in such chain.

2.19 "***Participant***" shall mean any Eligible Employee who has executed a subscription agreement and been granted rights to purchase Common Shares pursuant to the Plan.

2.20 "***Plan***" shall mean this 2018 Employee Share Purchase Plan.

2.21 "***Public Trading Date***" shall mean the first date upon which the Common Shares are listed (or approved for listing) upon notice of issuance on any securities exchange or designated (or approved for designation) upon notice of issuance as a national market security on an interdealer quotation system, or, if earlier, the date on which the Company becomes a "publicly held corporation" for purposes of Treasury Regulation Section 1.162-27(c)(1).

2.22 "***Purchase Date***" shall mean the last Trading Day of each Offering Period.

2.23 "***Purchase Price***" shall mean the purchase price designated by the Administrator in the applicable Offering Document (which purchase price shall not be less than 85% of the Fair Market Value of a Share on the Enrollment Date or on the Purchase Date, whichever is lower); provided, however, that, in the event no purchase price is designated by the Administrator in the applicable Offering Document, the purchase price for the Offering Periods

covered by such Offering Document shall be 85% of the Fair Market Value of a Share on the Enrollment Date or on the Purchase Date, whichever is lower; provided, further, that the Purchase Price may be adjusted by the Administrator pursuant to Article VIII and shall not be less than the par value of a Share.

2.24 "***Securities Act***" shall mean the Securities Act of 1933, as amended.

2.25 "***Share***" shall mean a Common Share.

2.26 "***Subject Person***" shall have the meaning given to such term in Section 2.4.

2.27 "***Subsidiary***" shall mean any corporation, other than the Company, in an unbroken chain of corporations beginning with the Company if, at the time of the determination, each of the corporations other than the last corporation in an unbroken chain owns shares possessing 50% or more of the total combined voting power of all classes of shares in one of the other corporations in such chain; provided, however, that a limited liability company or partnership may be treated as a Subsidiary to the extent either (a) such entity is treated as a disregarded entity under Treasury Regulation Section 301.7701-3(a) by reason of the Company or any other Subsidiary that is a corporation being the sole owner of such entity, or (b) such entity elects to be classified as a corporation under Treasury Regulation Section 301.7701-3(a) and such entity would otherwise qualify as a Subsidiary.

2.25 "***Trading Day***" shall mean a day on which national stock exchanges in the United States are open for trading.

ARTICLE III.
SHARES SUBJECT TO THE PLAN

3.1 Number of Shares. Subject to Article VIII, the aggregate number of Shares that may be issued pursuant to rights granted under the Plan shall be 670,000 Shares. In addition to the foregoing, subject to Article VIII, on the first day of each calendar year beginning on January 1, 2019 and ending on and including January 1, 2028, the number of Shares available for issuance under the Plan shall be increased by that number of Shares equal to the lesser of (a) 1% of the Shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (b) such smaller number of Shares as determined by the Board. If any right granted under the Plan shall for any reason terminate without having been exercised, the Common Shares not purchased under such right shall again become available for issuance under the Plan. Notwithstanding anything in this Section 3.1 to the contrary, the number of Shares that may be issued or transferred pursuant to the rights granted under the Plan shall not exceed an aggregate of 6,420,000 Shares, subject to Article VIII.

3.2 Shares Distributed. Any Common Shares distributed pursuant to the Plan may consist, in whole or in part, of authorized and unissued Common Shares, Common Shares held in treasury or Common Shares purchased on the open market.

ARTICLE IV.
OFFERING PERIODS; OFFERING DOCUMENTS; PURCHASE DATES

4.1 Offering Periods. The Administrator may from time to time grant or provide for the grant of rights to purchase Common Shares under the Plan to Eligible Employees during one or more periods (each, an "***Offering Period***") selected by the Administrator. The terms and conditions applicable to each Offering Period shall be set forth in an "***Offering Document***" adopted by the Administrator, which Offering Document shall be in such form and shall contain such terms and conditions as the Administrator shall deem appropriate and shall be incorporated by reference into and made part of the Plan and shall be attached hereto as part of the Plan. The provisions of separate Offering Periods under the Plan need not be identical.

4.2 Offering Documents. Each Offering Document with respect to an Offering Period shall specify (through incorporation of the provisions of this Plan by reference or otherwise):

(a) the length of the Offering Period, which period shall not exceed twenty-seven months;

(b) the maximum number of Shares that may be purchased by any Eligible Employee during such Offering Period, which, in the absence of a contrary designation by the Administrator, shall be 25,000 Shares; and

(c) such other provisions as the Administrator determines are appropriate, subject to the Plan.

ARTICLE V.
ELIGIBILITY AND PARTICIPATION

5.1 Eligibility. Any Eligible Employee who shall be employed by the Company or a Designated Subsidiary on a given Enrollment Date for an Offering Period shall be eligible to participate in the Plan during such Offering Period, subject to the requirements of this Article V and the limitations imposed by Section 423(b) of the Code.

5.2 Enrollment in Plan.

(a) Except as otherwise set forth in an Offering Document or determined by the Administrator, an Eligible Employee may become a Participant in the Plan for an Offering Period by delivering a subscription agreement to the Company by such time prior to the Enrollment Date for such Offering Period (or such other date specified in the Offering Document) designated by the Administrator and in such form as the Company provides.

(b) Each subscription agreement shall designate a whole percentage of such Eligible Employee's Compensation to be withheld by the Company or the Designated Subsidiary employing such Eligible Employee on each payday during the Offering Period as payroll deductions under the Plan. The percentage of Compensation designated by an Eligible Employee may not be less than 1% and may not be more than the maximum percentage specified by the Administrator in the applicable Offering Document (which percentage shall be 25% in the absence of any such designation) as payroll deductions. The payroll deductions made for each Participant shall be credited to an account for such Participant under the Plan and shall be deposited with the general funds of the Company.

(c) A Participant may increase or decrease the percentage of Compensation designated in his or her subscription agreement, subject to the limits of this Section 5.2, or may suspend his or her payroll deductions, at any time during an Offering Period; provided, however, that the Administrator may limit the number of changes a Participant may make to his or her payroll deduction

elections during each Offering Period in the applicable Offering Document (and in the absence of any specific designation by the Administrator, a Participant shall be allowed one change to his or her payroll deduction elections during each Offering Period). Any such change or suspension of payroll deductions shall be effective with the first full payroll period following five business days after the Company's receipt of the new subscription agreement (or such shorter or longer period as may be specified by the Administrator in the applicable Offering Document). In the event a Participant suspends his or her payroll deductions, such Participant's cumulative payroll deductions prior to the suspension shall remain in his or her account and shall be applied to the purchase of Shares on the next occurring Purchase Date and shall not be paid to such Participant unless he or she withdraws from participation in the Plan pursuant to Article VII.

(d) Except as otherwise set forth in an Offering Document or determined by the Administrator, a Participant may participate in the Plan only by means of payroll deduction and may not make contributions by lump sum payment for any Offering Period.

5.3 Payroll Deductions. Except as otherwise provided in the applicable Offering Document, payroll deductions for a Participant shall commence on the first payroll following the Enrollment Date and shall end on the last payroll in the Offering Period to which the Participant's authorization is applicable, unless sooner terminated by the Participant as provided in Article VII or suspended by the Participant or the Administrator as provided in Section 5.2 and Section 5.6, respectively.

5.4 Effect of Enrollment. A Participant's completion of a subscription agreement will enroll such Participant in the Plan for each subsequent Offering Period on the terms contained therein until the Participant either submits a new subscription agreement, withdraws from participation under the Plan as provided in Article VII or otherwise becomes ineligible to participate in the Plan.

5.5 Limitation on Purchase of Common Shares. An Eligible Employee may be granted rights under the Plan only if such rights, together with any other rights granted to such Eligible Employee under "employee stock purchase plans" of the Company, any Parent or any Subsidiary, as specified by Section 423(b)(8) of the Code, do not permit such employee's rights to purchase shares of the Company or any Parent or Subsidiary to accrue at a rate that exceeds $25,000 of the fair market value of such shares (determined as of the first day of the Offering Period during which such rights are granted) for each calendar year in which such rights are outstanding at any time. This limitation shall be applied in accordance with Section 423(b)(8) of the Code.

5.6 Decrease or Suspension of Payroll Deductions. Notwithstanding the foregoing, to the extent necessary to comply with Section 423(b)(8) of the Code and Section 5.5 or the other limitations set forth in this Plan, a Participant's payroll deductions may be suspended by the Administrator at any time during an Offering Period. The balance of the amount credited to the account of each Participant that has not been applied to the purchase of Shares by reason of Section 423(b)(8) of the Code, Section 5.5 or the other limitations set forth in this Plan shall be paid to such Participant in one lump sum in cash as soon as reasonably practicable after the Purchase Date.

5.7 Non-U.S. Employees. In order to facilitate participation in the Plan, the Administrator may provide for such special terms applicable to Participants who are citizens or residents of a country other than the United States, or who are employed by a Designated Subsidiary outside of the United States, as the Administrator may consider necessary or appropriate to accommodate differences in local law, tax policy or custom. Such special terms may not be more favorable than the terms of rights granted under the Plan to Eligible Employees who are residents of the United States. Moreover, the Administrator may approve such supplements to, or amendments, restatements or alternative versions of, this Plan as it may consider necessary or appropriate for such purposes without thereby affecting the

terms of this Plan as in effect for any other purpose. No such special terms, supplements, amendments or restatements shall include any provisions that are inconsistent with the terms of this Plan as then in effect unless this Plan could have been amended to eliminate such inconsistency without further approval by the shareholders of the Company.

5.8 Leave of Absence. During leaves of absence approved by the Company meeting the requirements of Treasury Regulation Section 1.421-1(h)(2) under the Code, a Participant may continue participation in the Plan by making cash payments to the Company on his or her normal payday equal to his or her authorized payroll deduction.

ARTICLE VI.
GRANT AND EXERCISE OF RIGHTS

6.1 Grant of Rights. On the Enrollment Date of each Offering Period, each Eligible Employee participating in such Offering Period shall be granted a right to purchase the maximum number of Shares specified under Section 4.2, subject to the limits in Section 5.5, and shall have the right to buy, on each Purchase Date during such Offering Period (at the applicable Purchase Price), such number of whole Shares as is determined by dividing (a) such Participant's payroll deductions accumulated prior to such Purchase Date and retained in the Participant's account as of the Purchase Date, by (b) the applicable Purchase Price (rounded down to the nearest Share). The right shall expire on the last day of the Offering Period.

6.2 Exercise of Rights. On each Purchase Date, each Participant's accumulated payroll deductions and any other additional payments specifically provided for in the applicable Offering Document will be applied to the purchase of whole Shares, up to the maximum number of Shares permitted pursuant to the terms of the Plan and the applicable Offering Document, at the Purchase Price. No fractional Shares shall be issued upon the exercise of rights granted under the Plan, unless the Offering Document specifically provides otherwise. Any cash in lieu of fractional Shares remaining after the purchase of whole Shares upon exercise of a purchase right will be credited to a Participant's account and carried forward and applied toward the purchase of whole Shares for the next following Offering Period. Shares issued pursuant to the Plan may be evidenced in such manner as the Administrator may determine and may be issued in certificated form or issued pursuant to book-entry procedures.

6.3 Pro Rata Allocation of Shares. If the Administrator determines that, on a given Purchase Date, the number of Shares with respect to which rights are to be exercised may exceed (a) the number of Shares that were available for issuance under the Plan on the Enrollment Date of the applicable Offering Period, or (b) the number of Shares available for issuance under the Plan on such Purchase Date, the Administrator may in its sole discretion provide that the Company shall make a pro rata allocation of the Shares available for purchase on such Enrollment Date or Purchase Date, as applicable, in as uniform a manner as shall be practicable and as it shall determine in its sole discretion to be equitable among all Participants for whom rights to purchase Common Shares are to be exercised pursuant to this Article VI on such Purchase Date, and shall either (i) continue all Offering Periods then in effect, or (ii) terminate any or all Offering Periods then in effect pursuant to Article IX. The Company may make pro rata allocation of the Shares available on the Enrollment Date of any applicable Offering Period pursuant to the preceding sentence, notwithstanding any authorization of additional Shares for issuance under the Plan by the Company's shareholders subsequent to such Enrollment Date. The balance of the amount credited to the account of each Participant that has not been applied to the purchase of Shares shall be paid to such Participant in one lump sum in cash as soon as reasonably practicable after the Purchase Date.

6.4 Withholding. At the time a Participant's rights under the Plan are exercised, in whole or in part, or at the time some or all of the Common Shares issued under the Plan is disposed of, the Participant must make adequate provision for the Company's federal, state, or other tax withholding obligations, if any, that arise upon the exercise of the right or the disposition of the Common Shares. At any time, the Company may, but shall not be obligated to, withhold from the Participant's compensation the amount necessary for the Company to meet applicable withholding obligations, including any withholding required to make available to the Company any tax deductions or benefits attributable to sale or early disposition of Common Shares by the Participant.

6.5 Conditions to Issuance of Common Shares. The Company shall not be required to issue or deliver any certificate or certificates for, or make any book entries evidencing, Shares purchased upon the exercise of rights under the Plan prior to fulfillment of all of the following conditions:

(a) The admission of such Shares to listing on all stock exchanges, if any, on which the Common Shares are then listed;

(b) The completion of any registration or other qualification of such Shares under any state or federal law or under the rulings or regulations of the Securities and Exchange Commission or any other governmental regulatory body, that the Administrator shall, in its absolute discretion, deem necessary or advisable;

(c) The obtaining of any approval or other clearance from any state or federal governmental agency that the Administrator shall, in its absolute discretion, determine to be necessary or advisable;

(d) The payment to the Company of all amounts that it is required to withhold under federal, state or local law upon exercise of the rights, if any; and

(e) The lapse of such reasonable period of time following the exercise of the rights as the Administrator may from time to time establish for reasons of administrative convenience.

ARTICLE VII.
WITHDRAWAL; CESSATION OF ELIGIBILITY

7.1 Withdrawal. A Participant may withdraw all but not less than all of the payroll deductions credited to his or her account and not yet used to exercise his or her rights under the Plan at any time by giving written notice to the Company in a form acceptable to the Company no later than one week prior to the end of the Offering Period. All of the Participant's payroll deductions credited to his or her account during an Offering Period shall be paid to such Participant as soon as reasonably practicable after receipt of notice of withdrawal and such Participant's rights for the Offering Period shall be automatically terminated, and no further payroll deductions for the purchase of Shares shall be made for such Offering Period. If a Participant withdraws from an Offering Period, payroll deductions shall not resume at the beginning of the next Offering Period unless the Participant timely delivers to the Company a new subscription agreement.

7.2 Future Participation. A Participant's withdrawal from an Offering Period shall not have any effect upon his or her eligibility to participate in any similar plan that may hereafter be adopted by the Company or a Designated Subsidiary or in subsequent Offering Periods that commence after the termination of the Offering Period from which the Participant withdraws.

7.3 Cessation of Eligibility. Upon a Participant's ceasing to be an Eligible Employee for any reason, he or she shall be deemed to have elected to withdraw from the Plan pursuant to this Article VII and the payroll deductions credited to such Participant's account during the Offering Period shall be paid to such Participant or, in the case of his or her death, to the person or persons entitled thereto under Section 12.4, as soon as reasonably practicable, and such Participant's rights for the Offering Period shall be automatically terminated.

ARTICLE VIII.
ADJUSTMENTS UPON CHANGES IN SHARES

8.1 Changes in Capitalization. Subject to Section 8.3, in the event that the Administrator determines that any dividend or other distribution (whether in the form of cash, Common Shares, other securities, or other property), Change in Control, reorganization, merger, amalgamation, consolidation, combination, repurchase, recapitalization, liquidation, dissolution, or sale, transfer, exchange or other disposition of all or substantially all of the assets of the Company, or sale or exchange of Common Shares or other securities of the Company, issuance of warrants or other rights to purchase Common Shares or other securities of the Company, or other similar corporate transaction or event, as determined by the Administrator, affects the Common Shares such that an adjustment is determined by the Administrator to be appropriate in order to prevent dilution or enlargement of the benefits or potential benefits intended by the Company to be made available under the Plan or with respect to any outstanding purchase rights under the Plan, the Administrator shall make equitable adjustments, if any, to reflect such change with respect to (a) the aggregate number and type of Shares (or other securities or property) that may be issued under the Plan (including, but not limited to, adjustments of the limitations in Section 3.1 and the limitations established in each Offering Document pursuant to Section 4.2 on the maximum number of Shares that may be purchased); (b) the class(es) and number of Shares and price per Share subject to outstanding rights; and (c) the Purchase Price with respect to any outstanding rights.

8.2 Other Adjustments. Subject to Section 8.3, in the event of any transaction or event described in Section 8.1 or any unusual or nonrecurring transactions or events affecting the Company, any affiliate of the Company, or the financial statements of the Company or any affiliate (including without limitation any Change in Control), or of changes in Applicable Law or accounting principles, the Administrator, in its discretion, and on such terms and conditions as it deems appropriate, is hereby authorized to take any one or more of the following actions whenever the Administrator determines that such action is appropriate in order to prevent the dilution or enlargement of the benefits or potential benefits intended to be made available under the Plan or with respect to any right under the Plan, to facilitate such transactions or events or to give effect to such changes in laws, regulations or principles:

(a) To provide for either (i) termination of any outstanding right in exchange for an amount of cash, if any, equal to the amount that would have been obtained upon the exercise of such right had such right been currently exercisable or (ii) the replacement of such outstanding right with other rights or property selected by the Administrator in its sole discretion;

(b) To provide that the outstanding rights under the Plan shall be assumed by the successor or survivor corporation, or a parent or subsidiary thereof, or shall be substituted for by similar rights covering the shares of the successor or survivor corporation, or a parent or subsidiary thereof, with appropriate adjustments as to the number and kind of shares and prices;

(c) To make adjustments in the number and type of Shares (or other securities or property) subject to outstanding rights under the Plan and/or in the terms and conditions of outstanding rights and rights that may be granted in the future;

(d) To provide that Participants' accumulated payroll deductions may be used to purchase Common Shares prior to the next occurring Purchase Date on such date as the Administrator determines in its sole discretion and the Participants' rights under the ongoing Offering Period(s) shall be terminated; and

(e) To provide that all outstanding rights shall terminate without being exercised.

8.3 No Adjustment Under Certain Circumstances. No adjustment or action described in this Article VIII or in any other provision of the Plan shall be authorized to the extent that such adjustment or action would cause the Plan to fail to satisfy the requirements of Section 423 of the Code.

8.4 No Other Rights. Except as expressly provided in the Plan, no Participant shall have any rights by reason of any subdivision or consolidation of shares of shares of any class, the payment of any dividend, any increase or decrease in the number of shares of shares of any class or any dissolution, liquidation, merger, amalgamation or consolidation of the Company or any other corporation. Except as expressly provided in the Plan or pursuant to action of the Administrator under the Plan, no issuance by the Company of shares of shares of any class, or securities convertible into shares of shares of any class, shall affect, and no adjustment by reason thereof shall be made with respect to, the number of Shares subject to outstanding rights under the Plan or the Purchase Price with respect to any outstanding rights.

ARTICLE IX.
AMENDMENT, MODIFICATION AND TERMINATION

9.1 Amendment, Modification and Termination. The Administrator may amend, suspend or terminate the Plan at any time and from time to time; provided, however, that approval of the Company's shareholders shall be required to amend the Plan to: (a) increase the aggregate number, or change the type, of shares that may be sold pursuant to rights under the Plan under Section 3.1 (other than an adjustment as provided by Article VIII); (b) change the corporations or classes of corporations whose employees may be granted rights under the Plan; or (c) change the Plan in any manner that would cause the Plan to no longer be an "employee stock purchase plan" within the meaning of Section 423(b) of the Code.

9.2 Certain Changes to Plan. Without shareholder consent and without regard to whether any Participant rights may be considered to have been adversely affected, to the extent permitted by Section 423 of the Code, the Administrator shall be entitled to change the Offering Periods, limit the frequency and/or number of changes in the amount withheld from Compensation during an Offering Period, establish the exchange ratio applicable to amounts withheld in a currency other than U.S. dollars, permit payroll withholding in excess of the amount designated by a Participant in order to adjust for delays or mistakes in the Company's processing of withholding elections, establish

reasonable waiting and adjustment periods and/or accounting and crediting procedures to ensure that amounts applied toward the purchase of Common Shares for each Participant properly correspond with amounts withheld from the Participant's Compensation, and establish such other limitations or procedures as the Administrator determines in its sole discretion to be advisable that are consistent with the Plan.

9.3 Actions In the Event of Unfavorable Financial Accounting Consequences. In the event the Administrator determines that the ongoing operation of the Plan may result in unfavorable financial accounting consequences, the Administrator may, in its discretion and, to the extent necessary or desirable, modify or amend the Plan to reduce or eliminate such accounting consequence including, but not limited to:

(a) altering the Purchase Price for any Offering Period including an Offering Period underway at the time of the change in Purchase Price;

(b) shortening any Offering Period so that the Offering Period ends on a new Purchase Date, including an Offering Period underway at the time of the Administrator action; and

(c) allocating Shares.

Such modifications or amendments shall not require shareholder approval or the consent of any Participant.

9.4 Payments Upon Termination of Plan. Upon termination of the Plan, the balance in each Participant's Plan account shall be refunded as soon as practicable after such termination, without any interest thereon.

ARTICLE X.
TERM OF PLAN

The Plan shall be effective on the Effective Date. The effectiveness of the Plan shall be subject to approval of the Plan by the shareholders of the Company within twelve months following the date the Plan is first approved by the Board. No right may be granted under the Plan prior to such shareholder approval. No rights may be granted under the Plan during any period of suspension of the Plan or after termination of the Plan.

ARTICLE XI.
ADMINISTRATION

11.1 Administrator. Unless otherwise determined by the Board, the Administrator of the Plan shall be the Compensation Committee of the Board (or another committee or a subcommittee of the Board to which the Board delegates administration of the Plan) (such committee, the "***Committee***"). The Board may at any time vest in the Board any authority or duties for administration of the Plan.

11.2 Action by the Administrator. Unless otherwise established by the Board or in any charter of the Administrator, a majority of the Administrator shall constitute a quorum. The acts of a majority of the members present at any meeting at which a quorum is present and, subject to Applicable Law and the Bye-laws of the Company, acts approved in writing by a majority of the Administrator in lieu of a meeting, shall be deemed the acts of the Administrator. Each member of the Administrator is entitled to, in good faith, rely or act upon any report or other information furnished to that member by any officer or other employee of the Company or any Designated Subsidiary, the Company's independent certified public accountants, or any executive compensation consultant or other professional retained by the Company to assist in the administration of the Plan.

11.3 Authority of Administrator. The Administrator shall have the power, subject to, and within the limitations of, the express provisions of the Plan and Applicable Laws:

(a) To determine when and how rights to purchase Common Shares shall be granted and the provisions of each offering of such rights (which need not be identical).

(b) To designate from time to time which Subsidiaries of the Company shall be Designated Subsidiaries, which designation may be made without the approval of the shareholders of the Company.

(c) To construe and interpret the Plan and rights granted under it, and to establish, amend and revoke rules and regulations for its administration. The Administrator, in the exercise of this power, may correct any defect, omission or inconsistency in the Plan, in a manner and to the extent it shall deem necessary or expedient to make the Plan fully effective.

(d) To amend, suspend or terminate the Plan as provided in Article IX.

(e) Generally, to exercise such powers and to perform such acts as the Administrator deems necessary or expedient to promote the best interests of the Company and its Subsidiaries and to carry out the intent that the Plan be treated as an "employee stock purchase plan" within the meaning of Section 423 of the Code.

11.4 Decisions Binding. The Administrator's interpretation of the Plan, any rights granted pursuant to the Plan, any subscription agreement and all decisions and determinations by the Administrator with respect to the Plan are final, binding, and conclusive on all parties.

ARTICLE XII.
MISCELLANEOUS

12.1 Restriction upon Assignment. A right granted under the Plan shall not be transferable other than by will or the applicable laws of descent and distribution, and is exercisable during the Participant's lifetime only by the Participant. Except as provided in Section 12.4 hereof, a right under the Plan may not be exercised to any extent except by the Participant. The Company shall not recognize and shall be under no duty to recognize any assignment or alienation of the Participant's interest in the Plan, the Participant's rights under the Plan or any rights thereunder.

12.2 Rights as a Shareholder. With respect to Shares subject to a right granted under the Plan, a Participant shall not be deemed to be a shareholder of the Company, and the Participant shall not have any of the rights or privileges of a shareholder, until such Shares have been issued to the Participant or his or her nominee following exercise of the Participant's rights under the Plan. No adjustments shall be made for dividends (ordinary or extraordinary, whether in cash securities, or other property) or distribution or other rights for which the record date occurs prior to the date of such issuance, except as otherwise expressly provided herein or as determined by the Administrator.

12.3 Interest. No interest shall accrue on the payroll deductions or contributions of a Participant under the Plan.

12.4 Designation of Beneficiary.

(a) A Participant may, in the manner determined by the Administrator, file a written designation of a beneficiary who is to receive any Shares and/or cash, if any, from the Participant's account under the Plan in the event of such Participant's death subsequent to a Purchase Date on which the Participant's rights are exercised but prior to delivery to such Participant of such Shares and cash. In addition, a Participant may file a written designation of a beneficiary who is to receive any cash from the Participant's account under the Plan in the event of such Participant's death prior to exercise of the Participant's rights under the Plan. If the Participant is married and resides in a community property state, a designation of a person other than the Participant's spouse as his or her beneficiary shall not be effective without the prior written consent of the Participant's spouse.

(b) Such designation of beneficiary may be changed by the Participant at any time by written notice to the Company. In the event of the death of a Participant and in the absence of a

beneficiary validly designated under the Plan who is living at the time of such Participant's death, the Company shall deliver such Shares and/or cash to the executor or administrator of the estate of the Participant, or if no such executor or administrator has been appointed (to the knowledge of the Company), the Company, in its discretion, may deliver such Shares and/or cash to the spouse or to any one or more dependents or relatives of the Participant, or if no spouse, dependent or relative is known to the Company, then to such other person as the Company may designate.

12.5 Notices. All notices or other communications by a Participant to the Company under or in connection with the Plan shall be deemed to have been duly given when received in the form specified by the Company at the location, or by the person, designated by the Company for the receipt thereof.

12.6 Equal Rights and Privileges. Subject to Section 5.7, all Eligible Employees will have equal rights and privileges under this Plan so that this Plan qualifies as an "employee stock purchase plan" within the meaning of Section 423 of the Code. Subject to Section 5.7, any provision of this Plan that is inconsistent with Section 423 of the Code will, without further act or amendment by the Company, the Board or the Administrator, be reformed to comply with the equal rights and privileges requirement of Section 423 of the Code.

12.7 Use of Funds. All payroll deductions received or held by the Company under the Plan may be used by the Company for any corporate purpose, and the Company shall not be obligated to segregate such payroll deductions.

12.8 Reports. Statements of account shall be given to Participants at least annually, which statements shall set forth the amounts of payroll deductions, the Purchase Price, the number of Shares purchased and the remaining cash balance, if any.

12.9 No Employment Rights. Nothing in the Plan shall be construed to give any person (including any Eligible Employee or Participant) the right to remain in the employ of the Company or any Parent or Subsidiary or affect the right of the Company or any Parent or Subsidiary to terminate the employment of any person (including any Eligible Employee or Participant) at any time, with or without cause.

12.10 Notice of Disposition of Shares. Each Participant shall give prompt notice to the Company of any disposition or other transfer of any Shares purchased upon exercise of a right under the Plan if such disposition or transfer is made: (a) within two years from the Enrollment Date of the Offering Period in which the Shares were purchased or (b) within one year after the Purchase Date on which such Shares were purchased. Such notice shall specify the date of such disposition or other transfer and the amount realized, in cash, other property, assumption of indebtedness or other consideration, by the Participant in such disposition or other transfer.

12.11 Governing Law. The Plan and any agreements hereunder shall be administered, interpreted and enforced under the internal laws of Delaware without regard to conflicts of laws thereof or of any other jurisdiction.

12.12 Electronic Forms. To the extent permitted by Applicable Law and in the discretion of the Administrator, an Eligible Employee may submit any form or notice as set forth herein by means of an electronic form approved by the Administrator. Before the commencement of an Offering Period, the Administrator shall prescribe the time limits within which any such electronic form shall be submitted to the Administrator with respect to such Offering Period in order to be a valid election.

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KINIKSA PHARMACEUTICALS, LTD.

NON-EMPLOYEE DIRECTOR COMPENSATION PROGRAM

Non-employee members of the board of directors (the "***Board***") of Kiniksa Pharmaceuticals, Ltd. (the "***Company***") shall receive cash and equity compensation as set forth in this Non-Employee Director Compensation Program (this "***Program***"). The cash and equity compensation described in this Program shall be paid or be made, as applicable, automatically and without further action of the Board, to each member of the Board who is not an employee of the Company or any parent or subsidiary of the Company (each, a "***Non-Employee Director***") who is entitled to receive such cash or equity compensation, unless such Non-Employee Director declines the receipt of such cash or equity compensation by written notice to the Company. This Program shall remain in effect until it is revised or rescinded by further action of the Board. This Program may be amended, modified or terminated by the Board at any time in its sole discretion. The terms and conditions of this Program shall supersede any prior cash and/or equity compensation arrangements for service as a member of the Board between the Company and any of its Non-Employee Directors. No Non-Employee Director shall have any rights hereunder. This Program shall become effective on the date of the effectiveness of the Company's Registration Statement on Form S-1 relating to the initial public offering of common shares (the "***Effective Date***").

I. CASH COMPENSATION

A. Annual Retainers. Each Non-Employee Director shall receive an annual retainer of $35,000 for service on the Board.

B. Additional Annual Retainers. In addition, each Non-Employee Director shall receive the following annual retainers:

1. *Chairman of the Board or Lead Independent Director*. A Non-Employee Director serving as Chairman of the Board or Lead Independent Director shall receive an additional annual retainer of $22,500 for such service.

2. *Audit Committee*. A Non-Employee Director serving as Chairperson of the Audit Committee shall receive an additional annual retainer of $15,000 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Audit Committee shall receive an additional annual retainer of $7,500 for such service.

3. *Compensation Committee*. A Non-Employee Director serving as Chairperson of the Compensation Committee shall receive an additional annual retainer of $10,000 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Compensation Committee shall receive an additional annual retainer of $5,000 for such service.

4. *Nominating and Corporate Governance Committee*. A Non-Employee Director serving as Chairperson of the Nominating and Corporate Governance

Committee shall receive an additional annual retainer of $8,000 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Nominating and Corporate Governance Committee shall receive an additional annual retainer of $4,000 for such service.

C. Payment of Retainers. The annual retainers described in Sections I(A) and I(B) shall be earned on a quarterly basis based on a calendar quarter and shall be paid in cash by the Company in arrears not later than the fifteenth day following the end of each calendar quarter. In the event a Non-Employee Director does not serve as a Non-Employee Director, or in the applicable positions described in Section I(B), for an entire calendar quarter, the retainer paid to such Non-Employee Director shall be prorated for the portion of such calendar quarter actually served as a Non-Employee Director, or in such position, as applicable.

II. EQUITY COMPENSATION

Non-Employee Directors shall be granted the equity awards described below. The awards described below shall be granted under and shall be subject to the terms and provisions of the Company's 2018 Incentive Award Plan or any other applicable Company equity incentive plan then-maintained by the Company (the "***Equity Plan***") and shall be granted subject to award agreements, including attached exhibits, in substantially the form previously approved by the Board. All applicable terms of the Equity Plan apply to this Program as if fully set forth herein, and all grants of share options hereby are subject in all respects to the terms of the Equity Plan and the applicable award agreement.

A. Initial Awards. Each Non-Employee Director who is initially elected or appointed to the Board after the Effective Date shall receive an option to purchase 37,965 of the Company's Class A common shares on the date of such initial election or appointment. The awards described in this Section II(A) shall be referred to as "***Initial Awards***." No Non-Employee Director shall be granted more than one Initial Award.

B. Subsequent Awards. A Non-Employee Director who (i) has been serving as a Non-Employee Director on the Board for at least six (6) months as of the date of any annual meeting of the Company's shareholders after the Effective Date and (ii) will continue to serve as a Non-Employee Director immediately following such meeting, shall be automatically granted an option to purchase 18,760 of the Company's Class A common shares on the date of such annual meeting. The awards described in this Section II(B) shall be referred to as "***Subsequent Awards***." For the avoidance of doubt, a Non-Employee Director elected for the first time to the Board at an annual meeting of the Company's shareholders shall only receive an Initial Award in connection with such election, and shall not receive any Subsequent Award on the date of such meeting as well.

C. Termination of Employment of Employee Directors. Members of the Board who are employees of the Company or any parent or subsidiary of the Company who subsequently terminate their employment with the Company and any parent or subsidiary of the Company and remain on the Board will not receive an Initial Award pursuant to Section II(A) above, but to the extent that they are otherwise entitled, will receive, after termination of

employment with the Company and any parent or subsidiary of the Company, Subsequent Awards as described in Section II(B) above.

D. Terms of Awards Granted to Non-Employee Directors

1. *Exercise Price*. The per share exercise price of each option granted to a Non-Employee Director shall equal the Fair Market Value (as defined in the Equity Plan) of a share of the Company's Class A common shares on the date the option is granted.

2. *Vesting*. Each Initial Award shall vest and become exercisable as to one-third of the shares subject to the Initial Award on the first anniversary of the date of grant and as to the remainder in twenty-four (24) substantially equal monthly installments thereafter, such that the Initial Award shall be fully vested on the third anniversary of the date of grant, subject to the Non-Employee Director continuing in service as a Non-Employee Director through each such vesting date. Each Subsequent Award shall vest and become exercisable in twelve (12) substantially equal monthly installments following the date of grant, subject to the Non-Employee Director continuing in service on the Board as a Non-Employee Director through each such vesting date. Unless the Board otherwise determines, any portion of an Initial Award or Subsequent Award which is unvested or unexercisable at the time of a Non-Employee Director's termination of service on the Board as a Non-Employee Director shall be immediately forfeited upon such termination of service and shall not thereafter become vested and exercisable. All of a Non-Employee Director's Initial Awards and Subsequent Awards shall vest in full immediately prior to the occurrence of a Change in Control (as defined in the Equity Plan), to the extent outstanding at such time.

3. *Term*. The maximum term of each share option granted to a Non-Employee Director hereunder shall be ten (10) years from the date the option is granted.

* * * * *

Exhibit 10.16

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Thomas W. Beetham (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee is currently serving as the Company's Executive Vice President, Chief Legal Officer and Corporate Development and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the "Original Agreement"); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Founder Invention and Non-Disclosure Agreement" shall mean the Founder Invention and Non-Disclosure Agreement between the Employee and Company dated as of September 18, 2015.

(g) "Founder Non-Competition and Non-Solicitation Agreement" shall mean the Founder Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of September 18, 2015.

(h) "Good Reason" shall mean any of the following, occurring without the Employee's written consent:

(i) a demotion of the Employee to a position with responsibilities substantially less than the Employee's prior position; provided that, a change in title, reporting relationships and/or responsibilities of the Employee could, but do not necessarily in and of themselves, individually or in the aggregate, constitute a demotion for purposes of this Section 1(h)(i); and in all instances, the determination of whether a demotion has occurred shall be made by the Company in good faith; or

(ii) a requirement that the Employee relocate Employee's primary reporting location to a location more than fifty (50) miles from the location of the Company's current offices in Lexington, Massachusetts as of the Effective Date; or

(iii) a reduction of more than five percent (5%) of Employee's Base Salary, other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated to Employee; or

(iv) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b), 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith that are remedied by the Company promptly after receipt of notice thereof given by the Employee; or

(v) a breach by the Company of this Agreement.

(i) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(j) "Parent Board" shall mean the board of directors of the Parent.

(k) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its Executive Vice President, Chief Legal Officer and Corporate Development, and shall initially report to the Chief Executive Officer of the Company. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties

and responsibilities hereunder. During the Term, the Employee shall not engage in any other business activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $375,000, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with an initial target level of 35% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee for Good Reason. The Employee may terminate Employee's employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating Employee's employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

(f) By the Employee Other than for Good Reason. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(f), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b), 5(d) or 5(e) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below) and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Founder Invention and Non-Disclosure Agreement and the Founder Invention and Non-Disclosure Agreement, both of which the Company and Employee acknowledge and agree shall be considered separate contracts. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the

reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company. The parties acknowledge that the Employee is also indemnified by the Parent to the extent set forth in the Indemnification Agreement between the Parent and Employee dated December 16, 2015.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement and the Founder Non-Solicitation and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior

negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that the Employee holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE

Name: Thomas W. Beetham

KINIKSA PHARMACEUTICALS CORP.

By:
Name: Sanj K. Patel
Title: Chief Executive Officer

Exhibit 10.17

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Christopher Heberlig (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee is serving as the Company's Executive Vice President, Chief Financial Officer and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the "Original Agreement"); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

EVP (Founder)

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Founder Invention and Non-Disclosure Agreement" shall mean the Founder Invention and Non-Disclosure Agreement between the Employee and Company dated as of September 18, 2015.

(g) "Founder Non-Competition and Non-Solicitation Agreement" shall mean the Founder Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of September 18, 2015.

(h) "Good Reason" shall mean any of the following, occurring without the Employee's written consent:

(i) a demotion of the Employee to a position with responsibilities substantially less than the Employee's prior position; provided that, a change in title, reporting relationships and/or responsibilities of the Employee could, but do not necessarily in and of themselves, individually or in the aggregate, constitute a demotion for purposes of this Section 1(h)(i); and in all instances, the determination of whether a demotion has occurred shall be made by the Company in good faith; or

(ii) a requirement that the Employee relocate Employee's primary reporting location to a location more than fifty (50) miles from the location of the Company's current offices in Lexington, Massachusetts as of the Effective Date; or

(iii) a reduction of more than five percent (5%) of Employee's Base Salary, other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated to Employee; or

(iv) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b), 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith that are remedied by the Company promptly after receipt of notice thereof given by the Employee; or

(v) a breach by the Company of this Agreement.

(i) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(j) "Parent Board" shall mean the board of directors of the Parent.

(k) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its Executive Vice President, Chief Financial Officer, and shall initially report to the Chief Executive Officer of the Company. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business

activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $350,000, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with an initial target level of 35% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee for Good Reason. The Employee may terminate Employee's employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating Employee's employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

(f) By the Employee Other than for Good Reason. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(f), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b), 5(d) or 5(e) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below) and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Founder Invention and Non-Disclosure Agreement and the Founder Invention and Non-Disclosure Agreement, both of which the Company and Employee acknowledge and agree shall be considered separate contracts. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the

reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company. The parties acknowledge that the Employee is also indemnified by the Parent to the extent set forth in the Indemnification Agreement between the Parent and Employee dated December 16, 2015.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement and the Founder Non-Solicitation and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior

negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that the Employee holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, , may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE

Name: Christopher Heberlig

KINIKSA PHARMACEUTICALS CORP.

By:
Name: Sanj K. Patel
Title: Chief Executive Officer

Exhibit 10.18

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Carsten Boess (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee is currently serving as the Company's Executive Vice President, Corporate Affairs and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the "Original Agreement");

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

EVP (Founder)

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Founder Invention and Non-Disclosure Agreement" shall mean the Founder Invention and Non-Disclosure Agreement between the Employee and Company dated as of September 18, 2015.

(g) "Founder Non-Competition and Non-Solicitation Agreement" shall mean the Founder Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of September 18, 2015.

(h) "Good Reason" shall mean any of the following, occurring without the Employee's written consent:

(i) a demotion of the Employee to a position with responsibilities substantially less than the Employee's prior position; provided that, a change in title, reporting relationships and/or responsibilities of the Employee could, but do not necessarily in and of themselves, individually or in the aggregate, constitute a demotion for purposes of this Section 1(h)(i); and in all instances, the determination of whether a demotion has occurred shall be made by the Company in good faith; or

(ii) a requirement that the Employee relocate Employee's primary reporting location to a location more than fifty (50) miles from the location of the Company's current offices in Lexington, Massachusetts as of the Effective Date; or

(iii) a reduction of more than five percent (5%) of Employee's Base Salary, other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated to Employee; or

(iv) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b), 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith that are remedied by the Company promptly after receipt of notice thereof given by the Employee; or

(v) a breach by the Company of this Agreement.

(i) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(j) "Parent Board" shall mean the board of directors of the Parent.

(k) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its Exective Vice President, Corporate Affairs, and shall initially report to the Chief Executive Officer of the Company. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business

activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $400,000, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with an initial target level of 35% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee for Good Reason. The Employee may terminate Employee's employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating Employee's employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

(f) By the Employee Other than for Good Reason. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(f), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b), 5(d) or 5(e) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below) and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Founder Invention and Non-Disclosure Agreement and the Founder Invention and Non-Disclosure Agreement, both of which the Company and Employee acknowledge and agree shall be considered separate contracts. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the

reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company. The parties acknowledge that the Employee is also indemnified by the Parent to the extent set forth in the Indemnification Agreement between the Parent and Employee dated December 16, 2015.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement and the Founder Non-Solicitation and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior

negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that the Employee holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE

Name: Carsten Boess

KINIKSA PHARMACEUTICALS CORP.

By:
Name: Sanj K. Patel
Title: Chief Executive Officer

Exhibit 10.19

AMENDED AND RESTATED EMPLOYMENT AGREEMENT

This Amended and Restated Employment Agreement (this "Agreement") is made and entered into as of , 2018 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Rasmus Holm-Jorgensen (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee is currently serving as the Company's Senior Vice President, Chief Strategy and Portfolio Officer and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the "Original Agreement"); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

SVP (Founder)

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital stock by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Founder Invention and Non-Disclosure Agreement" shall mean the Founder Invention and Non-Disclosure Agreement between the Employee and Company dated as of September 18, 2015.

(g) "Founder Non-Competition and Non-Solicitation Agreement" shall mean the Founder Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of September 18, 2015.

(h) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(i) "Parent Board" shall mean the board of directors of the Parent.

(j) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its Senior Vice President, Chief Strategy and Portfolio Officer, and shall initially report to the Chief Operating Officer. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $337,740, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with an initial target level of 30% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year

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immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(e), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but un-reimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b) or 5(d) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary divided by twelve (12), then multiplied by the number of months of the Severance Period (as defined below) (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below), and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the

Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement). The "Severance Period" shall be nine (9) months; provided, that if the Employee's separation from service occurs during the twelve (12) months following a Change in Control, then the Severance Period shall be twelve (12) months.

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Founder Invention and Non-Disclosure Agreement and the Founder Invention and Non-Disclosure Agreement, both of which the Company and Employee acknowledge and agree shall be considered separate contracts. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited

to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of

Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to

a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement and the Founder Non-Solicitation and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that the Employee holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE

Name: Rasmus Holm-Jorgensen

KINIKSA PHARMACEUTICALS CORP.

By:
Name: Sanj K. Patel
Title: Chief Executive Officer

Exhibit 23.1

CONSENT OF INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM

We hereby consent to the use in this Amendment No. 1 to the Registration Statement on Form S-1 of Kiniksa Pharmaceuticals, Ltd. of our report dated February 27, 2018, except for the effects of the reverse share split discussed in Note 14 to the consolidated financial statements, as to which the date is May 14, 2018, relating to the financial statements of Kiniksa Pharmaceuticals, Ltd., which appears in this Registration Statement. We also consent to the reference to us under the heading "Experts" in such Registration Statement.

/s/ PricewaterhouseCoopers LLP

Boston, Massachusetts
May 14, 2018