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As confidentially submitted to the Securities and Exchange Commission on February 27, 2018.

Registration No. 333-

UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
Washington, D.C. 20549

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.
15 Walnut Street
Wellesley Hills, MA 02481
(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** **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	Proposed Maximum Aggregate Offering Price[1]	Amount of Registration Fee[2]
Class A Common Shares, par value $0.0001 per share	$	$

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

(2) Calculated pursuant to Rule 457(o) based on an estimate of the proposed maximum aggregate offering price.

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.

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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 , 2018

PRELIMINARY PROSPECTUS

Shares

KINIKSA PHARMACEUTICALS

Class A Common Shares

This is an initial public offering of our Class A common shares. All 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 $ and $. We intend to apply 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 11 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 191 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 % of our aggregate voting power and the holders of Class B common shares will account for the remaining % 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 of our Class A common shares for a period of 30 days after the date of this prospectus.

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**

Wedbush PacGrow

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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 acquiring, discovering, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have built 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 believe each of our product candidates has the potential to be the best-in-class or the first-approved treatment for its respective targeted indication.

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 diseases and that each represents a potential "pipeline-within-a-molecule."

Our Programs

The following table summarizes our current pipeline of product candidates:

Program & Target	Originator	Targeted Indication	Preclinical	Phase 1	Phase 2	Phase 3	Status and Anticipated Next Milestone	Commercial Rights
ARCALYST IL-1α & IL-1β	**Regeneron**	Recurrent Pericarditis					• Phase 2 clinical trial enrolling with interim data expected in 2018 • Commence Phase 3 clinical trial in 2018	• Worldwide (excl. Middle East and North Africa)
Mavrilimumab GM-CSFRα	**MedImmune**	Giant Cell Arteritis					• Commence Phase 2 clinical trial in 2018	• Worldwide
KPL-716 OSMRβ	**Biogen**	Prurigo Nodularis / Atopic Dermatitis					• Enrolling Phase 1a / 1b clinical trial in healthy volunteers and subjects with atopic dermatitis • Interim Phase 1a / 1b data expected in 2018	• Worldwide
KPL-045 CD30L	**Novo Nordisk**	Autoimmune					• IND-filing planned for 2019	• Worldwide
KPL-404 CD40	**Primatope**	Autoimmune					• IND-filing planned for 2019	• Option for Worldwide
Discovery	Internal	Autoimmune					• Target / drug discovery efforts	• Worldwide

Note: ARCALYST is approved and marketed for cryopyrin-associated periodic syndrome, or CAPS, in the United States by Regeneron. We will assume the rights to this indication upon receiving regulatory approval for ARCALYST in recurrent pericarditis or a second indication.

- **ARCALYST** (rilonacept) represents a potentially best-in-class 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. We are initially developing ARCALYST for the treatment of recurrent pericarditis, a debilitating inflammatory cardiovascular disease. We are not aware of any therapy currently approved by the U.S. Food and Drug Administration, or the FDA, for the treatment of recurrent pericarditis. We are conducting an open-label Phase 2 proof-of-concept clinical trial in this disease, and we expect to report interim data from this trial in 2018. If the results from this trial are favorable, we plan to initiate a Phase 3 clinical trial in 2018.

- **Mavrilimumab** is a monoclonal antibody that antagonizes the signaling of granulocyte-macrophage colony growth factor, or GM-CSF. We believe it has the potential to be a best-in-class treatment of giant cell arteritis, or GCA, an inflammatory disease of the blood vessels with high unmet medical need. We plan to initiate a Phase 2 clinical trial of mavrilimumab for the treatment of GCA in 2018.

- **KPL-716** represents a highly-differentiated, potentially best-in-class monoclonal antibody for a variety of pruritic and fibrotic indications driven by the cytokines interleukin-31, or IL-31, and oncostatin M, or OSM, by simultaneously inhibiting both pathways from signaling through their common receptor subunit, oncostatin M receptor beta, or OSMRb. We believe KPL-716 is the only monoclonal antibody in development that simultaneously targets both pathways. We are currently enrolling subjects in a hybrid Phase 1a/1b clinical trial in healthy volunteers and in subjects with atopic dermatitis as a proof-of-concept for pruritic conditions. We expect to report interim data from this trial in 2018. If the data from this 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 Investigational New Drug, or IND, enabling studies 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 and expect to file an IND with the FDA for this program in 2019.

In addition to the indications described above, we plan to evaluate ARCALYST, 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 acquiring, discovering, 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 quickly as possible. We have a rigorous clinical development program with well-defined clinical milestones across our pipeline, which we believe provide near-and long-term catalysts for value growth. In 2018, we expect to report interim Phase 2 data for ARCALYST and, if the Phase 2 data are favorable, we plan to initiate a Phase 3 clinical trial for ARCALYST in recurrent pericarditis. In 2018, we also expect to initiate a Phase 2 clinical trial of mavrilimumab in GCA and to report interim Phase 1a/1b data for KPL-716.

- **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 have the potential to be a "pipeline-within-a-molecule." 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, ARCALYST, 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, ARCALYST, 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 % of our aggregate voting power and the Class B common shares will account for the remaining % of the aggregate voting power. In addition, the number of Class A1 common shares to be outstanding after this offering will be and the number of Class B1 common shares to be outstanding after this offering will be . See "Principal Shareholders" and "Description of Share Capital" for more information on beneficial ownership immediately following this offering.

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, and those third parties may not perform satisfactorily, which could delay our product development activities;
- 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	shares
Option to purchase additional Class A common shares	shares
Class A common shares to be outstanding after this offering	shares (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	shares
Class A1 common shares to be outstanding after this offering	shares
Class B1 common shares to be outstanding after this offering	shares
Total common shares to be outstanding after this offering	shares (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 % of our aggregate voting power and the holders of our Class B common shares will account for the remaining % 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 $ million (or approximately $ 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 $ 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 for the clinical and pre-clinical development of our product candidates, working capital and general corporate purposes. See "Use of Proceeds" beginning on page 80.
Risk factors	See "Risk Factors" beginning on page 11 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 Class A common shares and Class B common shares outstanding as of , 2018 and assumes the conversion of all of our preferred shares outstanding as of , 2018 into Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares, in each case upon the closing of this offering. This amount excludes:

- Class A common shares issuable upon exercise of share options outstanding as of , 2018, at a weighted average exercise price of $ per share;
- Class A common shares reserved for future issuance under our 2015 Equity Incentive Plan as of , 2018; and
- Class A common shares that will become available for future issuance under our 2018 Equity Incentive Plan, which will become effective in connection with this offering upon the effectiveness of the registration statement of which this prospectus forms a part.

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

- a 1-for reverse stock split of our common shares effected on , 2018;
- the conversion of all of our preferred shares into Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares, in each case upon the closing of this offering;
- no exercise of outstanding share options after , 2018;
- the effectiveness of our amended and restated bye-laws immediately prior to the closing of this offering; and
- no exercise by the underwriters of their option to purchase additional Class A common shares.

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 and the consolidated balance sheet data as of December 31, 2017 from our audited consolidated financial statements included elsewhere in this prospectus. Our historical results are not necessarily indicative of the results that should be expected in the future.

	Year Ended December 31,	
	2016	2017
	(in thousands, except share and per share data)	
Consolidated Statement of Operations Data:		
Operating expenses:		
Research and development	$ 17,439	$ 56,357
General and administrative	6,563	9,043
Total operating expenses	24,002	65,400
Loss from operations	(24,002)	(65,400)
Interest income	65	529
Loss before provision for income taxes	(23,937)	(64,871)
Provision for income taxes	(36)	(2)
Net loss	$ (23,973)	$ (64,873)
Net loss per share attributable to common shareholders—basic and diluted[(1)]	$ (33.53)	$ (13.12)
Weighted average common shares outstanding—basic and diluted[(1)]	715,045	4,944,889
Pro forma net loss per share attributable to common shareholders—basic and diluted (unaudited)[(1)]		$ (1.00)
Pro forma weighted average common shares outstanding—basic and diluted (unaudited)[(1)]		65,588,468

(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. The pro forma net loss per share attributable to common shareholders presented in this table does not give effect to the sale and issuance of our Series C preferred shares in February 2018.

	As of December 31, 2017		
	Actual	Pro Forma[2]	Pro Forma As Adjusted[3]
	(in thousands)		
Consolidated Balance Sheet Data:			
Cash and cash equivalents	$ 45,555	$	$
Working capital[1]	29,674		
Total assets	47,492		
Convertible preferred shares	119,770		
Total shareholders' equity (deficit)	(89,708)		

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

(2) The pro forma balance sheet data give effect to (i) the sale and issuance of 34,932,049 shares of our Series C preferred shares in February 2018 for aggregate gross proceeds of $200.0 million and (ii) the automatic conversion of all of our outstanding preferred shares into an aggregate of common shares upon closing of this offering.

(3) The pro forma as adjusted balance sheet data give further effect to our issuance and sale of Class A common shares in this offering at an assumed initial public offering price of $ 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 $ 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 $ 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 $ 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 were $24.0 million and $64.9 million, respectively. As of December 31, 2017, we had an accumulated deficit of $91.0 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 ARCALYST 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 ARCALYST 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, ARCALYST, 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 candidiates 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 ARCALYST for the treatment of recurrent pericarditis and our Phase 1 clinical trials of KPL-716 for the treatment of atopic dermatitis and commencing our planned Phase 3 clinical trial for ARCALYST 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 ARCALYST 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 ARCALYST, 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, and 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 ARCALYST is approved and marketed for human use for the treatment of CAPS in the United States by Regeneron, we are studying ARCALYST for the treatment of recurrent pericarditis in an open-label Phase 2 proof-of-concept clinical trial, and, if the data is favorable, 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 is 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, and we have yet to generate clinical data of the products we are studying for the indications we are pursuing. 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 data from our ongoing open-label Phase 2 proof-of-concept clinical trial of ARCALYST, we plan to initiate a Phase 3 clinical trial for ARCALYST 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 ARCALYST 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 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, 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, such as for mavrilimumab or KPL-716, 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 ARCALYST 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 ARCALYST, 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 ARCALYST 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 evaluated 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. 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 "top-line" 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 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 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, 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.

ARCALYST 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 ARCALYST, 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 ARCALYST 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 ARCALYST, 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 ARCALYST, 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 ARCALYST, 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 ARCALYST 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 ARCALYST, 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 ARCALYST, mavrilimumab and KPL-716 are supplied to us from single-source suppliers. For example, although Regeneron has been producing ARCALYST 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 ARCALYST, 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 ARCALYST, 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, which is expected to enter early-stage clinical trials in GCA in 2018. In addition, AbbVie is conducting clinical trials for an oral janus kinase inhibitor, and Sanofi S.A. and Regeneron intend to initiate a Phase 3 trial for their anti- interleukin-6 program 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 products currently in development for atopic dermatitis including upadacitinib, PF-04965842, ANB-020, nemolizumab, baracitinib, 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.

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 ARCALYST, 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 ARCALYST, 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 ARCALYST, 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 ARCALYST 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 ARCALYST 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 ARCALYST 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 ARCALYST 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, ARCALYST, 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 ARCALYST. 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 ARCALYST 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 ARCALYST in fields to which we have licensed the rights, but we retain the commercial benefit related to that development upon approval of ARCALYST 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 ARCALYST in CAPS and certain periodic fever syndromes. The development of ARCALYST in other fields could increase the possibility of identification of adverse safety results that impact our development of ARCALYST for recurrent pericarditis. In addition, if approved, commercialization of ARCALYST in other fields could result in an increased threat of off-label use to compete with the sale of ARCALYST to treat these indications, which may diminish sales of ARCALYST 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 ARCALYST 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. Kiniksa is 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, members of our senior management team and entities affiliated with certain of our directors 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, members of our senior management team and entities affiliated with certain of our directors will hold % 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. These shareholders 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. These shareholders 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, as a result of such conversion, the holder and its affiliates would own more than 9.99% of the combined voting power of our share capital outstanding unless such holders provide us with 61-days' prior notice that they intend to increase their ownership of our voting share capital above 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.

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 senior management team 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 , 2018, upon the completion of this offering, we will have outstanding a total of Class A common shares, Class A1 common shares, Class B common shares and 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 , 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 shares of Class A common shares will be eligible for sale in the public market. Approximately 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 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, 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 our Class A 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 $ per Class A common share, which is the midpoint of the price range set forth on the cover page of this prospectus, you will experience immediate dilution of $ 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 % of the aggregate price paid by all purchasers of our common shares but will own only approximately % 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 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 a specified election once 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 Class A common shares in this offering will be approximately $ million (or $ million if the underwriters exercise in full their option to purchase additional Class A common shares), assuming an initial public offering price of $ 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 $ 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 $ 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 $ 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 of this offering, together with our existing cash and cash equivalents, to fund our clinical and pre-clinical development programs and the remainder for working capital and other general corporate purposes.

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 enable us to fund our operating expenses and capital expenditure requirements through . 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.

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 prior to 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 December 31, 2017:

- on an actual basis;
- on a pro forma basis to give effect to:
  - the issuance and sale of 34,932,049 Series C preferred shares in February 2018 for aggregate gross proceeds of $200.0 million;
  - the automatic conversion of all outstanding preferred shares into Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares upon the closing of this offering; and
  - the effectiveness of our amended and restated bye-laws immediately prior to the closing of this offering; and
- on a pro forma as adjusted basis to give further effect to our issuance and sale of Class A common shares in this offering at an assumed initial public offering price of $ 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 December 31, 2017		
	Actual	Pro Forma	Pro Forma As Adjusted
	(in thousands, except share and per share data)		
Cash and cash equivalents	$ 45,555	$	$
Convertible preferred shares (Series A, B and C), par value of $0.0001 per share; 62,531,219 shares designated, issued and outstanding, actual; and no shares designated, issued and outstanding, pro forma and pro forma as adjusted	$ 119,770	$	$
Shareholders' equity (deficit):			
Class A common shares, $0.0001 par value; 15,049,615 shares designated, 1,967,242 shares issued and outstanding, actual; shares designated, shares issued and outstanding, pro forma; shares designated, shares issued and outstanding, pro forma as adjusted	—		
Class B common shares, $0.0001 par value; 9,750,005 shares designated, issued and outstanding, actual; shares designated, shares issued and outstanding, pro forma; shares designated, shares issued and outstanding, pro forma as adjusted	1		
Class A1 common shares, $0.0001 par value; no shares designated, issued and outstanding, actual; shares designated, shares issued and outstanding, pro forma; shares designated, shares issued and outstanding, pro forma as adjusted	—		
Class B1 common shares, $0.0001 par value; no shares designated, issued and outstanding, actual; shares designated, shares issued and outstanding, pro forma; shares designated, shares issued and outstanding, pro forma as adjusted	—		
Additional paid-in capital	1,289		
Accumulated deficit	(90,998)		
Total shareholders' equity (deficit)	(89,708)		
Total capitalization	$ 30,062	$	$

Each $1.00 increase (decrease) in the assumed initial public offering price of $ 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 $ 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 $ 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:

- 8,533,432 Class A common shares issuable upon exercise of share options outstanding as of December 31, 2017, at a weighted average exercise price of $1.01 per share;
- 4,548,941 Class A common shares reserved for future issuance under our 2015 Equity Incentive Plan as of December 31, 2017; and
- 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 upon the effectiveness of the registration statement of which this prospectus forms a part.

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 December 31, 2017 was $(89.7) million, or $(7.66) 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 11,717,247 common shares outstanding as of December 31, 2017.

Our pro forma net tangible book value as of December 31, 2017 was $ million, or $ 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 (i) the issuance and sale of 34,932,049 Series C preferred shares in February 2018 for gross proceeds of $200.0 million and (ii) the automatic conversion of all outstanding preferred shares into an aggregate of 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 December 31, 2017, after giving effect to the pro forma adjustments described above.

After giving further effect to our issuance and sale of Class A common shares in this offering at an assumed initial public offering price of $ 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 December 31, 2017 would have been $ million, or $ per share. This represents an immediate increase in pro forma as adjusted net tangible book value of $ per share to our existing shareholders and immediate dilution of $ 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		$
Historical net tangible book value (deficit) per share as of December 31, 2017	$ (7.66)	
Increase per share attributable to the pro forma adjustments described above		
Pro forma net tangible book value per share as of December 31, 2017		
Increase in pro forma as adjusted net tangible book value per share attributable to new investors purchasing Class A common shares in this offering		
Pro forma as adjusted net tangible book value per share after this offering		
Dilution per share to new investors purchasing Class A common shares in this offering		$

Each $1.00 increase (decrease) in the assumed initial public offering price of $ 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 $ and the dilution per share to new investors by $, 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 $ and decrease the dilution per share to new investors by $, 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 $ and increase the dilution per share to new investors by $, 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 $, and the dilution per share to new investors would be $, in each case assuming an initial public offering price of $ 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 $ 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		%	$	%	$
New investors					
Total		100.0%	$	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 % 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 % of the total number of our common shares outstanding after this offering.

The foregoing tables exclude:

- 8,533,432 Class A common shares issuable upon exercise of share options outstanding as of December 31, 2017, at a weighted average exercise price of $1.01 per share;
- 4,548,941 Class A common shares reserved for future issuance under our 2015 Equity Incentive Plan as of December 31, 2017; and
- Class A common shares that will become available for future issuance under our 2018 Equity Incentive Plan, which will become effective in connection with this offering.

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.

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. Our historical results are not necessarily indicative of the results that should be expected in the future.

	Year Ended December 31,	
	2016	2017
	(in thousands, except share and per share data)	
Consolidated Statement of Operations Data:		
Operating expenses:		
Research and development	$ 17,439	$ 56,357
General and administrative	6,563	9,043
Total operating expenses	24,002	65,400
Loss from operations	(24,002)	(65,400)
Interest income	65	529
Loss before provision for income taxes	(23,937)	(64,871)
Provision for income taxes	(36)	(2)
Net loss	$ (23,973)	$ (64,873)
Net loss per share attributable to common shareholders—basic and diluted[(1)]	$ (33.53)	$ (13.12)
Weighted average common shares outstanding—basic and diluted[(1)]	715,045	4,944,889
Pro forma net loss per share attributable to common shareholders—basic and diluted (unaudited)[(1)]		$ (1.00)
Pro forma weighted average common shares outstanding—basic and diluted (unaudited)[(1)]		65,588,468

(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. The pro forma net loss per share attributable to common shareholders presented in this table does not give effect to the sale and issuance of our Series C preferred shares in February 2018.

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

(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 acquiring, discovering, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have built 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 believe each of our product candidates has the potential to be the best-in-class or the first-approved treatment for its respective targeted indication.

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 diseases and that each represents a potential "pipeline-within-a-molecule."

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 December 31, 2017, we had received net proceeds of $119.8 million from the sale of preferred shares. In February 2018, we received gross proceeds of $200.0 million from the sale of Series C 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. As of December 31, 2017, we had an accumulated deficit of $91.0 million. 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, we had cash and cash equivalents of $45.6 million. In February 2018, we received gross proceeds of $200.0 million in connection with our closing of our Series C preferred share financing. 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,	
	2016	2017
	(in thousands)	
ARCALYST[1]	$ —	$ 6,301
Mavrilimumab[2]	—	18,000
KPL-716[3]	14,870	24,164
KPL-045[4]	—	1,654
KPL-404[5]	—	549
Unallocated research and development expenses	2,569	5,689
Total research and development expenses	$ 17,439	$ 56,357

(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.

(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 ARCALYST, 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 profile with IND-enabling 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 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:			
ARCALYST	$ —	$ 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 ARCALYST 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 ARCALYST 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 1 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 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 partnering with CMOs on development and manufacturing of drug supply for our product candidates and partnering with CROs on the conduct and oversight of our Phase 1 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 ARCALYST program. Personnel-related costs for the years ended December 31, 2016 and 2017 included share-based compensation of $0.1 million and $0.3 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, 2016 and 2017 included share-based compensation of $0.3 million and $0.6 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, 2016 and 2017.

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 December 31, 2017, we had received net proceeds of $119.8 million from sales of our preferred shares. In February 2018, we received gross proceeds of $200.0 million from the issuance and sale of our Series C preferred shares. As of December 31, 2017, we had cash and cash equivalents of $45.6 million.

Cash Flows

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

	Year Ended December 31,	
	2016	2017
	(in thousands)	
Net cash used in operating activities	$ (21,867)	$ (50,219)
Net cash used in investing activities	(3)	(69)
Net cash provided by financing activities	42,509	39,873
Net increase (decrease) in cash and cash equivalents and restricted cash	$ 20,639	$ (10,415)

Operating Activities

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 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 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 ARCALYST, 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 ARCALYST 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.

(3) Represents minimum payments due for our lease of office space in Wellesley Hills, Massachusetts under an operating lease agreement that expires in August 2018.

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 ARCALYST 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 of low double-digit percentages on annual net sales of licensed products. 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 a specified 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 of high single-digit to low double-digit percentages on annual net sales of products licensed under the agreement.

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 $0.58 per share as of October 15, 2015, $0.68 per share as of September 16, 2016, $1.39 per share as of March 8, 2017 and June 29, 2017, $1.63 per share as of September 4, 2017 and $1.71 as of December 14, 2017.

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	75,000	$ 0.58	$ 0.58	$ 0.38
June 1, 2016	61,000	$ 0.58	$ 0.58	$ 0.35
September 14, 2016	673,312	$ 0.68	$ 0.68	$ 0.43
December 7, 2016	91,500	$ 0.68	$ 0.68	$ 0.44
March 6, 2017	107,500	$ 1.39	$ 1.39	$ 0.93
June 7, 2017	117,000	$ 1.39	$ 1.39	$ 0.91
June 29, 2017	3,484,186	$ 1.39	$ 1.39	$ 0.92
September 14, 2017	215,000	$ 1.63	$ 1.63	$ 1.07
December 14, 2017	303,000	$ 1.71	$ 1.71	$ 1.12

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 and December 31, 2017, 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 10% change 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 acquiring, discovering, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have built 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 believe each of our product candidates has the potential to be the best-in-class or the first-approved treatment for its respective targeted indication.

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 diseases and that each represents a potential "pipeline-within-a-molecule."

The following table summarizes our current pipeline of product candidates:

Program & Target	Originator	Targeted Indication	Preclinical	Phase 1	Phase 2	Phase 3	Status and Anticipated Next Milestone	Commercial Rights
ARCALYST IL-1α & IL-1β	Regeneron	Recurrent Pericarditis					• Phase 2 clinical trial enrolling with interim data expected in 2018 • Commence Phase 3 clinical trial in 2018	• Worldwide (excl. Middle East and North Africa)
Mavrilimumab GM-CSFRα	MedImmune	Giant Cell Arteritis					• Commence Phase 2 clinical trial in 2018	• Worldwide
KPL-716 OSMRβ	Biogen	Prurigo Nodularis / Atopic Dermatitis					• Enrolling Phase 1a / 1b clinical trial in healthy volunteers and subjects with atopic dermatitis • Interim Phase 1a / 1b data expected in 2018	• Worldwide
KPL-045 CD30L	Novo Nordisk	Autoimmune					• IND-filing planned for 2019	• Worldwide
KPL-404 CD40	Primatope	Autoimmune					• IND-filing planned for 2019	• Option for Worldwide
Discovery	Internal	Autoimmune					• Target / drug discovery efforts	• Worldwide

Note: ARCALYST is approved and marketed for cryopyrin-associated periodic syndrome, or CAPS, in the United States by Regeneron. We will assume the rights to this indication upon receiving regulatory approval for ARCALYST in recurrent pericarditis or a second indication.

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.

- **ARCALYST** (rilonacept) represents a potentially best-in-class 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. We are initially developing ARCALYST for the treatment of recurrent pericarditis, a debilitating inflammatory cardiovascular disease. We are not aware of any therapy currently approved by the U.S. Food and Drug Administration, or the FDA, for the treatment of recurrent pericarditis. We are conducting an open-label Phase 2 proof-of-concept clinical trial in this disease, and we expect to report interim data from this trial in 2018. If the results from this trial are favorable, we plan to initiate a Phase 3 clinical trial in 2018.

- **Mavrilimumab** is a monoclonal antibody that antagonizes the signaling of granulocyte-macrophage colony growth factor, or GM-CSF. We believe it has the potential to be a best-in-class treatment of giant cell arteritis, or GCA, an inflammatory disease of the blood

vessels with high unmet medical need. We plan to initiate a Phase 2 clinical trial of mavrilimumab for the treatment of GCA in 2018.

- **KPL-716** represents a highly-differentiated, potentially best-in-class monoclonal antibody for a variety of pruritic and fibrotic indications driven by the cytokines interleukin-31, or IL-31, and oncostatin M, or OSM, by simultaneously inhibiting both pathways from signaling through their common receptor subunit, oncostatin M receptor beta, or OSMRb. We believe KPL-716 is the only monoclonal antibody in development that simultaneously targets both pathways. We are currently enrolling subjects in a hybrid Phase 1a/1b clinical trial in healthy volunteers and in subjects with atopic dermatitis as a proof-of-concept for pruritic conditions. We expect to report interim data from this trial in 2018. If the interim data from this 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 Investigational New Drug, or IND, enabling studies 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 and expect to file an IND with the FDA for this program in 2019.

In addition to the indications described above, we plan to evaluate ARCALYST, 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 acquiring, discovering, 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. We have a rigorous clinical development program with well-defined clinical milestones across our pipeline, which we believe provide near-and long-term catalysts for value growth. In 2018, we expect to report interim Phase 2 data for ARCALYST and, if the Phase 2 data are favorable, we plan to initiate a Phase 3 clinical trial for ARCALYST in recurrent pericarditis. In 2018, we also expect to initiate a Phase 2 clinical trial of mavrilimumab in GCA and to report interim Phase 1a/1b data for KPL-716.

- **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 have the potential to be a "pipeline-within-a-molecule." 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, ARCALYST, 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, ARCALYST, 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

ARCALYST (rilonacept)

Overview

ARCALYST is an FDA-approved therapy for cryopyrin-associated periodic syndrome, or CAPS, marketed in the United States by Regeneron Pharmaceuticals, Inc., or Regeneron, and has potential to treat certain diseases mediated by both IL-1a and IL-1b. Our lead indication for ARCALYST 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 interim 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 ARCALYST in 2018. We also plan to evaluate ARCALYST 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 ARCALYST 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.

Before we licensed ARCALYST in 2017, Regeneron developed ARCALYST. In addition to CAPS, ARCALYST is approved in the United States for familial cold auto-inflammatory syndrome and Muckle-Wells syndrome and is marketed by Regeneron.

Mechanism of Action

ARCALYST 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 ARCALYST 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 ARCALYST 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 ARCALYST 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

ARCALYST is a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1a and IL-1b signaling. We believe ARCALYST has the potential to be a best-in-class and first-approved therapy for recurrent pericarditis. Beyond recurrent pericarditis, we believe there is significant potential for ARCALYST to address additional indications, including larger 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 ARCALYST 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 ARCALYST 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 c-reactive protein, or 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: Asymptomatic subjects with recurrent pericarditis receiving NSAIDS +/– colchicine +/– steroids who are dependent or unable to wean off their current therapy;
- Part 4: Symptomatic subjects with postpericardiotomy syndrome receiving NSAIDS +/– colchicine +/– steroids with high CRP measurements; and
- Part 5: Asymptomatic subjects with postpericardiotomy syndrome receiving NSAIDS +/– colchicine +/– steroids who are dependent or unable to wean off their current therapy.

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 11-point Numerical Rating Scale instrument for assessment of pericardial pain in subjects with symptomatic recurrent idiopathic pericarditis both at baseline and on treatment with ARCALYST 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 ARCALYST. This trial has been designed with the potential to examine the response of patients with pericarditis caused by a variety of underlying etiologies. We expect to report interim data from this trial in 2018. If those results are positive, we plan to initiate a Phase 3 clinical trial in 2018.

Clinical History of ARCALYST

Regeneron evaluated ARCALYST 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 ARCALYST 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 ARCALYST. In the pivotal efficacy trial, which evaluated the long-term efficacy and safety of once-weekly dosing, 160 mg of ARCALYST markedly decreased the clinical signs and symptoms of CAPS.

- Gout: Regeneron evaluated ARCALYST 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, ARCALYST 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, among other things, additional clinical data in order to further assess the risk-benefit profile of ARCALYST in treating gout.

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

In these trials, the most common adverse events reported were injection site reactions and upper respiratory tract infections. In the Phase 2 and Phase 3 programs for gout flare prevention, which treated the largest number of patients, the most common adverse events reported for the 160 mg dose of ARCALYST were injection site reactions (15.5% for ARCALYST versus 2.6% for placebo) and upper respiratory tract infections (10.3% for ARCALYST 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 contribute 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 successful, clinically-relevant and statistically-significant effect mavrilimumab had on primary and secondary efficacy measures in multiple Phase 2 trials conducted by MedImmune in patients with RA.

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 100,000 to 200,000 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 development of mavrilimumab in other vasculitides where GM-CSF signaling has been implicated.

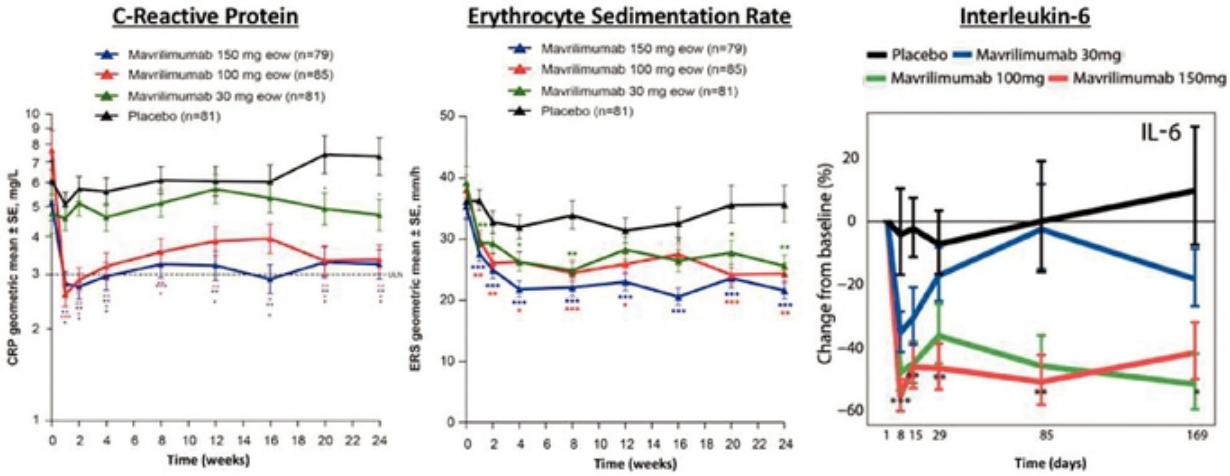
Clinical History in Rheumatoid Arthritis

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, leaving the clinical hold in place, and 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 potentially best-in-class, 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 hybrid Phase 1a/Phase 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 1 clinical trial are favorable, we plan to initiate further trials in prurigo nodularis and atopic dermatitis. We expect to report interim data from the Phase 1 clinical trials in 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-4R a, in human skin equivalent cultures, upregulates IL-4R a 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 18.0 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-4Ra) 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 interact 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 hybrid 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 Phase 1b portion of the trial, the cohorts of subjects with moderate to severe atopic dermatitis are receiving dose levels of KPL-716 which could show an early signal of efficacy in reducing pruritus as an exploratory endpoint using a validated Numerical Rating Score which measures pruritus intensity. In the first portion of the Phase 1b clinical trial, each subject will receive a single dose of KPL-716 administered intravenously. 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 interim data from the Phase 1a and the first portion of the Phase 1b (single-ascending dose) clinical trial in 2018.

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 conducting activities that we anticipate will allow us to submit an IND 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 conducting activities that we anticipate will allow us to submit an IND 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 ARCALYST (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 ARCALYST 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 ARCALYST 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 ARCALYST outside of the Excluded Indications in our territory. Upon receiving positive data in a Phase 3 clinical trial, Regeneron will transfer the BLA for ARCALYST 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 ARCALYST 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 ARCALYST. 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 an ARCALYST product developed by us. We may also terminate with three months' written notice if we reasonably determine that ARCALYST 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 ARCALYST for clinical development. If Regeneron determines to discontinue the supply of ARCALYST to us, it must use its reasonable efforts to transfer all relevant documentation, materials and technology necessary for the manufacture of ARCALYST 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 ARCALYST.

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 of low double-digit percentages on annual net sales of licensed products. 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 a specified anniversary of first commercial sale of such product in such country.

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 of high single-digit to low double-digit percentages on annual net sales of licensed products. We have also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to KPL-716.

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 ARCALYST 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 ARCALYST for recurrent pericarditis, we intend to market and commercialize ARCALYST 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 ARCALYST, 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 ARCALYST, 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:

ARCALYST

We are not aware of any therapies currently approved by the FDA for the treatment of recurrent pericarditis, our lead indication for ARCALYST. 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, AbbVie is conducting clinical trials for an oral janus kinase inhibitor, 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 Lily and Co., Pfizer Inc., AbbVie, Glenmark Pharmaceuticals Ltd., GSK, LEO Pharma Inc., Incyte Corporation, Dermavant Sciences, Inc. and AnaptysBio, 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 in the United States and numerous foreign jurisdictions relating to ARCALYST. A U.S. patent covering ARCALYST 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 ARCALYST 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. 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. 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 ARCALYST, 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 Investigational New Drug Application, or 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 December 31, 2017, we had 41 employees.

Facilities

Our offices are located in Wellesley Hills, Massachusetts, where we have leased approximately 10,800 square feet of office space, under a lease which expires in August 2018. 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 February 27, 2018.

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 Baker, Ph.D.	48	Director
Stephen R. Biggar, M.D., Ph.D.	47	Director
Thomas Malley	49	Director
Tracey McCain	50	Director
Kimberly Popovits	59	Director
Barry D. Quart, Pharm.D.	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 founded 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 2013, 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 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 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 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 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 upon 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 the holders of at least two-thirds of the votes that all our shareholders would be entitled to cast in an annual general election of directors, and that any vacancy on our board of directors, including a vacancy resulting from an enlargement of our board of directors, 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 upon the closing of this offering, 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. Upon the closing of this offering, the members of the classes will be divided as follows:

- the Class I directors will be and , and their terms will expire at our first annual meeting of shareholders following this offering;
- the Class II directors will be and , and their terms will expire at our second annual meeting of shareholders following this offering; and
- the Class III directors will be , and , and their terms will expire at the third annual meeting of shareholders following this offering.

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, , and 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 director is Felix 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 , and . 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 and meet the independence requirements of Rule 10A-3 under the Exchange Act and the applicable Nasdaq rules. Our board of directors has determined that 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 , and . serves as the chairperson of the committee. Our board of directors has determined that each of , and is independent under the applicable Nasdaq rules, including the Nasdaq rules specific to membership on the compensation committee and 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 , and . serves as the chairperson of the committee. Our board of directors has determined that , and 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 have generally 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.

The following table sets forth the option awards granted to our named executive officers in the 2017 fiscal year.

Named Executive Officer	2017 Option Awards Granted
Sanj K. Patel	704,863
Stephen Mahoney	240,032
John F. Paolini, M.D., Ph.D.	300,938

These options were issued under our 2015 Stock 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, we intend to adopt a 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. We expect that the 2018 Plan will be effective on the day prior to the first public trading date of our Class A common shares, subject to approval of such plan by our shareholders. For additional information about the 2018

Plan, please see the section titled "Executive Compensation Plans — 2018 Incentive Award Plan" 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	802,941	573,530	0.58	12/15/2025
	6/28/2017	—	704,863	1.39	6/28/2027
Stephen Mahoney	8/1/2015	200,735	143,382	0.58	12/15/2025
	6/28/2017	—	240,032	1.39	6/28/2027
John F. Paolini, M.D., Ph.D.	8/15/2016	207,271	414,541	0.68	9/13/2026
	6/28/2017	—	300,938	1.39	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 enter into new employment agreements with our named executive officers that will supersede their current employment agreements or to amend their existing employment agreements. The new or amended employment agreements will become effective upon the effectiveness of the registration statement of which this prospectus is a part. The material terms of those arrangements are not currently known and will be described in this prospectus once finally determined.

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.

We intend to approve and implement a new compensation program for our directors that will become effective on the effectiveness of the registration statement of which this prospectus is a part. The terms of this program are not yet known.

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 Baker, Ph.D.	—	—	—
Stephen R. Biggar, M.D., Ph.D.	—	—	—
Thomas 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 directors other than Mr. Patel who were serving 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 Baker, Ph.D.	—
Stephen R. Biggar, M.D., Ph.D.	—
Thomas Malley	135,000
Barry D. Quart, Pharm.D	135,000

Executive Compensation Plans

The following summarizes the material terms of the 2018 Plan, the long-term incentive compensation plan 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 13,099,614 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. 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 and which following the effective date of the 2018 Plan are not issued under the 2015 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 appropriately 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

Effective the day prior to the first public trading date of our Class A common shares, we intend to adopt and ask our shareholders to approve 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 determine which eligible service providers receive awards, grant 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 Class A common shares will initially be available for issuance under the 2018 Plan. No more than 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 will count against the maximum number of shares that may be issued upon the exercise of ISOs.

In addition, the maximum aggregate grant date fair value as determined in accordance with FASB ASC Topic 718 (or any successor thereto), of awards granted to any non-employee director for services as a director pursuant to the 2018 Plan during any fiscal year may not exceed $ (or, in the fiscal year of any director's initial service, $). The plan administrator may, however, make exceptions to such limit on director compensation in extraordinary circumstances, subject to the limitations in the 2018 Plan.

Awards

The 2018 Plan provides for the grant of options to purchase shares, including ISOs and options that are not intended to qualify as ISOs under the Code, non-qualified stock 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 Shares or Cash Based Awards.* Other shares 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 shares 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 shares 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. The 2018 Plan will remain in effect until the tenth anniversary of its effective date, 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.

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 our share capital 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.

Preferred Share Financings

Series A Preferred Share Financing

In October 2015, we issued and sold an aggregate of 21,937,521 Series A Preferred Shares, and in September 2016 we sold an additional 24,862,523 Series A Preferred Shares to new investors and certain of our directors and executive officers at a price of $1.7094 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 15,731,175 Series B Preferred Shares to new investors, existing investors, a director and an executive officer at a price of $2.5427 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 34,932,049 Series C Preferred Shares to new investors, existing investors and certain executive officers at a price of $5.7254 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 our share capital 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[2]	43,875,042	9,832,068	11,352,918
HH RSV-XVII Holdings Limited	—	3,146,261	8,733,014
Arrowpoint Funds	—	1,573,000	1,222,621
Executive Officers			
Sanj K. Patel[3]	1,170,001	196,641	174,760
Stephen Mahoney	292,500	—	17,466
Thomas Beetham	131,625	—	17,466
Chris Heberlig	204,750	—	17,466
Thomas Malley[4]	—	196,641	—

(1) Additional details regarding these shareholders and their equity holdings are provided under the caption "Principal Shareholders."

(2) 667, L.P. and Baker Brothers Life Sciences, L.P. participated in our preferred share financing. See "Principal Shareholders" for additional details.

(3) Additional details regarding this named executive officer's equity holdings are provided under the caption "Principal Shareholders."

(4) Mr. Malley's investment was made through his affiliated entity, Mossrock Capital, LLC.

Our directors, Felix 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 share capital.

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 our share capital 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 share capital and entities affiliated with certain of our directors. Pursuant to the voting agreement, the preferred shareholders agree to elect the following directors to serve as members on our board of directors: Sanj K. Patel, Felix Baker, Ph.D, Stephen R. Biggar, M.D., Ph.D, Barry D. Quart, Pharm.D., Thomas Malley, Tracey McCain and Kimberly Popvits. As of the date of this prospectus, each of these directors continues to serve on our board of directors. Pursuant to the voting agreement, Dr. Baker and Dr. 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 enter into new employment agreements with our named executive officers or amend their existing employment agreements. The material terms of those arrangements are not currently known and will be described in this prospectus once finally determined in "Executive and Director Compensation — Executive Compensation Arrangements."

Share Option Grants to Executive Officers and Directors

We have granted share options to our executive officers and certain of our directors as more fully described in the section entitled "Executive and Director Compensation."

Policies and Procedures for Related Person Transactions

Our board of directors will adopt a written related person transaction policy, to be effective upon the closing of this offering, 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 , 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 (by voting power);
- 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 as to which the individual or entity has sole or shared voting power or investment power. Applicable percentage ownership is based on Class A common shares outstanding, Class A1 common shares outstanding, Class B common shares outstanding, and Class B1 common shares, each as of , 2018 and assumes the conversion of all outstanding preferred shares into Class A common shares, Class A1 common shares, Class B common shares, and Class B1 common shares, assuming an initial public offering price of $ per share, which is the midpoint of the price range set forth on the cover page of this prospectus. 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 , 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., 15 Walnut Street, Wellesley Hills, Massachusetts 02481. 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.

	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:																		
Entities Managed by Baker Bros. Advisors LLP(1)																		
HH RSV-XVII Holdings Limited(2)																		
Arrowpoint Funds(3)																		
Named Executive Officers and Directors:																		
Sanj K. Patel(4)																		
Stephen Mahoney(5)																		
John F. Paolini, M.D., Ph.D.(6)																		
Felix Baker, Ph.D.(1)																		
Stephen R. Biggar, M.D., Ph.D.(1)																		
Thomas Malley(7)																		
Tracey McCain																		
Kimberly Popovits																		
Barry D. Quart, Pharm.D.(8)																		
All executive officers and directors as a group (11 persons)(9)																		

* Represents beneficial ownership less than 1%.

† 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, as a result of such conversion, the holder and its affiliates would own more than 9.99% of the combined voting power of our outstanding share capital. 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 9.99% of the voting power of our outstanding share capital, 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, as a result of such conversion, the holder and its affiliates would own more than 9.99% of the combined voting power of our outstanding share capital. 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 9.99% of the voting power of our outstanding share capital, in addition to any other Class A common shares or Class B common shares beneficially owned by such holder.

(1) Consists of (a) Class B common shares and Class B1 common shares following conversion of Series A preferred shares held directly by Baker Brothers Life Sciences, L.P. ("Life Sciences"), (b) Class A common shares and Class A1 common shares following conversion of Series B preferred shares and Series C preferred shares held directly by Life Sciences, (c) Class B common shares and Class B1 common shares following conversion of Series A preferred shares held directly by 667, L.P. ("667") and (d) Class A common shares and Class A1 common shares following conversion of Series B preferred shares and Series C preferred shares held directly by 667. Baker Bros. Advisors LP is the Investment Adviser for the Baker Funds.

(2) Consists of Class A common shares and Class A1 common shares following conversion of Series B preferred shares and Series C preferred shares held by HH RSV-XVII Holdings Limited.

(3) Consists of Class A common shares and Class A1 common shares following conversion of Series B preferred shares and Series C preferred shares held by Arrowpoint Funds.

(4) Consists of (i) Class A common shares, (ii) Class B common shares and (iii) Class A common shares which Mr. Patel has the right to acquire pursuant to outstanding share options which are or will be exercisable within 60 days of , 2018.

(5) Consists of (i) Class A common shares, (ii) Class B common shares and (iii) Class A common shares which Mr. Mahoney has the right to acquire pursuant to outstanding share options which are or will be exercisable within 60 days of , 2018.

(6) Consists of Class A common shares which Dr. Paolini has the right to acquire pursuant to outstanding share options which are or will be exercisable within 60 days of , 2018.

(7) Consists of (i) Class A common shares and (ii) Class A common shares which Mr. Malley has the right to acquire pursuant to outstanding share options which are or will be exercisable within 60 days of , 2018.

(8) Consists of Class A common shares which Dr. Quart has the right to acquire pursuant to outstanding share options which are or will be exercisable within 60 days of , 2018.

(9) Consists of (i) Class A common shares, (ii) Class A1 common shares, (iii) Class B common shares, (iv) Class B1 common shares and (v) Class A common shares which the executive officers and directors have the right to acquire pursuant to outstanding share options which are or will be exercisable within 60 days of , 2018.

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 shareholders have approved certain amendments to our bye-laws that will become effective upon 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 designated share capital will consist of Class A common shares, par value $0.0001 per share, Class B common shares, par value $0.0001 per share, Class A1 common shares, par value $0.0001 per share and Class B1 common shares, par value $0.0001 per share, and there will be Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares issued and outstanding.

Pursuant to our 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 common 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 February 15, 2018, there were 1,967,242 Class A common shares issued and outstanding. All Class A common shares are fully paid and non-assessable.

Class B Common Shares

As of February 15, 2018, there were 9,750,005 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 February 15, 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, as a result of such conversion, the holder and its affiliates would own more than 9.99% of the combined voting power of our share capital outstanding. A holder of Class A1 common shares may increase 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 February 15, 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, as a result of such conversion, the holder and its affiliates would own more than 9.99% of the combined voting power of our share capital outstanding. A holder of Class B1 common shares may increase 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 that will become effective upon the consummation of this offering, our board of directors is authorized to issue preference 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 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 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 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 % of our aggregate voting power and the holders of Class B common shares will account for the remaining % 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 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 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 five 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 in excess of 50% of the total issued 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 such number of directors as the board of directors or members 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. The initial terms of the Class I, Class II and Class III directors will expire in 2019, 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 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 bye-laws or Bermuda law that directors hold any of our shares. There is also no requirement in our 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 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 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 our bye-laws will 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. 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 Business Combinations

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. Our amended and restated bye-laws provide that the approval of a simple majority of shareholders voting at a meeting to approve the amalgamation or merger agreement shall be sufficient, and the quorum for such meeting shall be two or more persons holding or representing more than 50% of the issued 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 our issued and voting shares outstanding 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 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 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 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 bye-laws, preference 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. Our bye-laws will 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 bye-laws, our directors may be removed only for cause by resolution passed by such number of shareholders that collectively hold more than 50% of the issued shares entitled to vote on such resolution. Any vacancy on our board, including a vacancy resulting from an enlargement of our board, 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 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 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.

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 , which will also serve as transfer agent. The transfer agent's address is .

Listing

We intend to apply 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 Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares, assuming the issuance of Class A common shares offered by us in this offering, the automatic conversion of all our outstanding preferred shares into Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares upon the closing of this offering, and no exercise of options after , 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 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 approximately shares will be available for sale in the public market, subject in some cases to applicable volume limitations under Rule 144.

In addition, of the Class A common shares that were subject to share options outstanding as of , 2018, options to purchase Class A common shares were vested as of , 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 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 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 upon 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 (including modifications adopted pursuant to our bye-laws) 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 stockholdersif 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 directorsexcept 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 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

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. 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

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 (1) we cease to be a PFIC and the U.S. Holder has made a "deemed sale" election under the PFIC rules, or (2) the U.S. Holder makes a QEF Election (defined below) with respect to taxable years in which we are a PFIC. 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 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. 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. 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	
Wedbush Securities Inc.	
Total	

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 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 Class A common additional 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 intend to list our common shares 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 $. 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 $.

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, Bermuda 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 each of the two years in the period ended December 31, 2017 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 and in accordance with auditing standards generally accepted in the United States of America. 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

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,		Pro Forma December 31,
	2016	2017	2017
			(unaudited)
Assets			
Current assets:			
Cash and cash equivalents	$ 55,970	$ 45,555	$ 45,555
Restricted cash	105	105	105
Prepaid expenses and other current assets	259	1,444	1,444
Total current assets	56,334	47,104	47,104
Property and equipment, net	84	125	125
Deferred offering costs	8	25	25
Deferred tax assets	41	238	238
Total assets	$ 56,467	$ 47,492	$ 47,492
Liabilities, Convertible Preferred Shares and Shareholders' Equity (Deficit)			
Current liabilities:			
Accounts payable	$ 212	$ 1,218	$ 1,218
Accrued expenses	2,090	6,212	6,212
Accrued milestone	—	10,000	10,000
Total liabilities	2,302	17,430	17,430
Commitments and contingencies (Note 12)			
Convertible preferred shares (Series A and B), $0.0001 par value; 46,800,044 shares and 62,531,219 shares designated, issued and outstanding as of December 31, 2016 and 2017, respectively; aggregate liquidation preference of $80,000 and $120,000 as of December 31, 2016 and 2017, respectively; no shares issued or outstanding, pro forma as of December 31, 2017 (unaudited)	79,897	119,770	
Shareholders' equity (deficit):			
Class A common shares, $0.0001 par value; 12,273,501 and 15,049,615 shares designated as of December 31, 2016 and 2017, respectively; 1,967,242 shares issued and outstanding as of December 31, 2016 and 2017; issued and outstanding, pro forma, as of December 31, 2017 (unaudited)	—	—	
Class B common shares, $0.0001 par value; 9,750,005 shares designated, issued and outstanding as of December 31, 2016 and 2017; shares issued and outstanding, pro forma, as of December 31, 2017 (unaudited)	1	1	
Class A1 common shares, $0.0001 par value; no shares designated, issued or outstanding as of December 31, 2016 and 2017; shares designated, issued and outstanding, pro forma as of December 31, 2017 (unaudited)	—	—	
Class B1 common shares, $0.0001 par value; no shares designated, issued or outstanding as of December 31, 2016 and 2017; shares designated, issued and outstanding, pro forma as of December 31, 2017 (unaudited)	—	—	
Additional paid-in capital	392	1,289	
Accumulated deficit	(26,125)	(90,998)	(90,998)
Total shareholders' equity (deficit)	(25,732)	(89,708)	30,062
Total liabilities, convertible preferred shares and shareholders' equity (deficit)	$ 56,467	$ 47,492	$ 47,492

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,	
	2016	**2017**
Operating expenses:		
Research and development	$ 17,439	$ 56,357
General and administrative	6,563	9,043
Total operating expenses	24,002	65,400
Loss from operations	(24,002)	(65,400)
Interest income	65	529
Loss before provision for income taxes	(23,937)	(64,871)
Provision for income taxes	(36)	(2)
Net loss and comprehensive loss	$ (23,973)	$ (64,873)
Net loss per share attributable to common shareholders—basic and diluted	$ (33.53)	$ (13.12)
Weighted average common shares outstanding—basic and diluted	715,045	4,944,889
Pro forma net loss per share attributable to common shareholders—basic and diluted (unaudited)		$ (1.00)
Pro forma weighted average common shares outstanding—basic and diluted (unaudited)		64,588,468

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 and B)		Common Shares (Class A and B)		Additional Paid-In	Accumulated	Total Shareholders'
	Shares	Amount	Shares	Amount	Capital	Deficit	Deficit
Balances at December 31, 2015	21,937,521	$ 37,398	11,700,006	$ 1	$ 14	$ (2,152)	$ (2,137)
Issuance of Series A convertible preferred shares, net of issuance costs of $1	24,862,523	42,499	—	—	—	—	—
Exercise of options	—	—	17,241	—	10	—	10
Share-based compensation expense	—	—	—	—	368	—	368
Net loss	—	—	—	—	—	(23,973)	(23,973)
Balances at December 31, 2016	46,800,044	79,897	11,717,247	1	392	(26,125)	(25,732)
Issuance of Series B convertible preferred shares, net of issuance costs of $127	15,731,175	39,873	—	—	—	—	—
Share-based compensation expense	—	—	—	—	897	—	897
Net loss	—	—	—	—	—	(64,873)	(64,873)
Balances at December 31, 2017	62,531,219	$ 119,770	11,717,247	$ 1	$ 1,289	$ (90,998)	$ (89,708)

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,	
	2016	**2017**
Cash flows from operating activities:		
Net loss	$ (23,973)	$ (64,873)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	22	28
Share-based compensation expense	368	897
Deferred income taxes	(46)	(197)
Changes in operating assets and liabilities:		
Prepaid expenses and other current assets	(219)	(1,185)
Accounts payable	119	1,006
Accrued expenses	1,862	4,105
Accrued milestone	—	10,000
Net cash used in operating activities	(21,867)	(50,219)
Cash flows from investing activities:		
Purchases of property and equipment	(3)	(69)
Net cash used in investing activities	(3)	(69)
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
Proceeds from exercise of options	10	—
Net cash provided by financing activities	42,509	39,873
Net increase (decrease) in cash and cash equivalents and restricted cash	20,639	(10,415)
Cash and cash equivalents and restricted cash at beginning of year	35,436	56,075
Cash and cash equivalents and restricted cash at end of year	$ 56,075	$ 45,660
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	$ 8	$ 25

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 acquiring, discovering, 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 built 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, 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. In addition, as of December 31, 2017, the Company had an accumulated deficit of $90,998. The Company expects to continue to generate operating losses for the foreseeable future. As of February 27, 2018, the issuance date of these consolidated financial statements, the Company expects 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 14), will be sufficient to fund its operating expenses and capital expenditure requirements for at least twelve months from the issuance date of these 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 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

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)

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, 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 Pro Forma Information

The accompanying unaudited pro forma consolidated balance sheet as of December 31, 2017 has been prepared to give effect, upon the closing of a qualified IPO, to the conversion of (i) all outstanding Series A convertible preferred shares into Class B common shares and Class B1 common shares and (ii) all outstanding Series B convertible preferred shares into Class A common shares and Class A1 common shares as if the Company's proposed IPO had occurred on December 31, 2017.

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 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 Class B common shares and Class B1 common shares and (ii) all outstanding Series B convertible preferred shares into Class A common shares and Class A1

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)

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, 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, 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.

Restricted Cash

Restricted cash as of December 31, 2016 and 2017 consisted of cash held in a money market fund in connection with the Company's corporate credit cards. Restricted cash amounts have been classified as current assets based on the contractual release date of the restrictions.

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

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)

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.

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.

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.

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)

- 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.

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.

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)

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.

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.

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)

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 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

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)

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 shareholders for the years ended December 31, 2016 and 2017.

Recently Adopted Accounting Pronouncements

In January 2017, the Financial Accounting Standards Board ("FASB") issued Accounting Standards Update ("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 (see Note 9).

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

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)

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 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.

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 May 2017, the FASB issued 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,

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)

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 will adopt 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.

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 public entities 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 will adopt ASU 2017-09 as of the required effective date of January 1, 2018, and the adoption is not expected to have an impact on the Company's financial position, results of operations or cash flows.

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 public entities 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.

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 2014-09 such that the standard is effective for public entities for annual periods beginning after

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)

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 adoption of ASU 2014-09 is not expected to have an impact on the Company's consolidated financial statements as the Company does not currently have any revenue-generating arrangements; however, the adoption of this standard will impact the accounting for any future revenue transactions.

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

During the years ended December 31, 2016 and 2017, 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 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 year 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,	
	2016	2017
Furniture and fixtures	$ 14	$ 83
Computer hardware and software	9	9
Vehicles	85	85
	108	177
Less: Accumulated depreciation	(24)	(52)
	$ 84	$ 125

Depreciation expense for the years ended December 31, 2016 and 2017 was $22 and $28, respectively.

5. Accrued Expenses

Accrued expenses consisted of the following:

	December 31,	
	2016	2017
Accrued employee compensation and benefits	$ 986	$ 1,570
Accrued research and development expenses	979	3,905
Accrued legal and professional fees	122	688
Other	3	49
	$ 2,090	$ 6,212

6. Convertible Preferred Shares

As of December 31, 2016 and 2017, the Company's bye-laws, as amended and restated (the "Amended Bye-Laws"), authorized the Company to issue 62,531,219 convertible preferred shares with a par value of $0.0001 per share, of which 46,800,044 shares have been designated as Series A convertible preferred shares (the "Series A preferred shares") and 15,731,175 shares have been designated as Series B convertible preferred shares (the "Series B 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 the control of the Company. Therefore, the Series A and Series B preferred shares (collectively, the "Preferred Shares") are classified outside of shareholders' equity (deficit).

In October 2015, the Company issued and sold 21,937,521 Series A preferred shares at a price of $1.7094 per share (the "Series A Original Issue Price") for proceeds of $37,398, net of issuance costs of $102.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

In September 2016, the Company issued and sold an additional 24,862,523 Series A preferred shares at a price of $1.7094 per share for proceeds of $42,499, net of issuance costs of $1.

In March 2017, the Company issued and sold 15,731,175 Series B preferred shares at a price of $2.5427 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 34,932,049 Series C convertible preferred shares (the "Series C preferred shares") at a price of $5.7254 per share for gross proceeds of $200,000 (see Note 14).

As of each balance sheet date, the Preferred Shares consisted of the following:

	December 31, 2016				
	Preferred Shares Authorized	Preferred Shares Issued and Outstanding	Carrying Value	Liquidation Preference	Common Shares Issuable Upon Conversion
Series A preferred shares	46,800,044	46,800,044	$ 79,897	$ 80,000	46,800,044

	December 31, 2017				
	Preferred Shares Authorized	Preferred Shares Issued and Outstanding	Carrying Value	Liquidation Preference	Common Shares Issuable Upon Conversion
Series A preferred shares	46,800,044	46,800,044	$ 79,897	$ 80,000	46,800,044
Series B preferred shares	15,731,175	15,731,175	39,873	40,000	15,731,175
	62,531,219	62,531,219	$ 119,770	$ 120,000	62,531,219

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). Except as

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

provided by law or by the other provisions of the Company's 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 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.

The Series A Original Issue Price and Series A Conversion Price were equal to $1.7094 as of December 31, 2016 and 2017. The Series B Original Issue Price and Series B Conversion Price were equal to $2.5427 as of December 31, 2017. Such Series A and Series B Original Issue Prices and Series A and Series B 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 and Series B Conversion Prices are also subject to adjustments based on weighted-average anti-dilution provisions set forth in the Company's Amended Bye-Laws in the event that additional securities are issued at a purchase price less than the Series A Conversion Price and/or the Series B Conversion Price then in effect. As of December 31, 2016 and 2017, each Series A preferred share was convertible into one Class B common share, and, as of December 31, 2017, each Series B 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 $5.1282 per share (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 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

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

6. Convertible Preferred Shares (Continued)

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 preferred shares may elect to receive Class A1 common shares in lieu of Class A common shares.

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 or Series B Original Issue Price. Through December 31, 2016 and 2017, 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 or Series B 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 Company's 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 Company's Amended Bye-Laws authorized the Company to issue 120,000,000 total shares with a par value of $0.0001, of which 12,273,501 and 15,049,615 shares have been designated as Class A common shares as of December 31, 2016 and 2017, respectively, and 9,750,005 shares have been designated as Class B common shares as of December 31, 2016 and 2017. The remaining 51,176,450 and 32,669,162 shares that were not designated as common shares or Preferred Shares as of December 31, 2016 and 2017, 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. 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, 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 10,323,500 and 13,099,614 shares as of December 31, 2016 and 2017, respectively, of which 5,979,013 shares remained available for future grant as of December 31, 2016 and 4,548,941 shares remained available for future grant as of December 31, 2017.

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 all awards may not be greater than 10 years, with the exception of options granted to persons possessing more than 10% of the total combined voting power of all classes of shares, which 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 years for employees and non-employees with 25% of the option vesting on the first anniversary of the grant date and the remaining shares vesting monthly for three 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.

During the years ended December 31, 2016 and 2017, the Company granted options to purchase 865,812 and 4,221,686 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 and $876 during the years ended December 31, 2016 and 2017, respectively.

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 35,000 and 5,000 Class A common shares, respectively, to non-employees. The Company recorded share-based compensation expense for options granted to non-employees of $14 and $21 during the years ended December 31, 2016 and 2017, 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,	
	2016	**2017**
Risk-free interest rate	1.45%	1.99%
Expected term (in years)	6.25	6.25
Expected volatility	70.75%	74.18%
Expected dividend yield	0%	0%

The assumptions that the Company used to determine the grant-date fair value of options granted to non-employees were as follows, presented on a weighted-average basis:

	Year Ended December 31,	
	2016	**2017**
Risk-free interest rate	1.94%	2.49%
Expected term (in years)	10.00	10.00
Expected volatility	65.85%	78.28%
Expected dividend yield	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, 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:

	Number of Shares	Weighted Average Exercise Price	Weighted Average Remaining Contractual Term (in years)	Aggregate Intrinsic Value
Outstanding as of December 31, 2016	4,327,246	$ 0.60	9.10	$ 356
Granted	4,226,686	1.43		
Exercised	—	—		
Forfeited	(20,500)	1.37		
Outstanding as of December 31, 2017	8,533,432	$ 1.01	8.82	$ 6,010
Options exercisable as of December 31, 2017	2,304,642	$ 0.59	8.07	$ 2,580
Options unvested as of December 31, 2017	6,228,790	$ 1.16	9.09	$ 3,431

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 17,241 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.

The weighted-average grant-date fair value per share of options granted during the years ended December 31, 2016 and 2017 was $0.42 and $0.94, respectively.

The total fair value of options vested during the years ended December 31, 2016 and 2017 was $402 and $445, 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.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

8. Share-Based Compensation (Continued)

The following table summarizes restricted share activity for the year ended December 31, 2017:

	Class A		Class B	
	Number of Shares	Weighted Average Grant Date Fair Value	Number of Shares	Weighted Average Grant Date Fair Value
Unvested restricted shares outstanding as of December 31, 2016	1,340,626	$ 0.0001	6,906,254	$ 0.0001
Granted	—	—	—	—
Vested	(487,500)	0.0001	(2,437,501)	0.0001
Unvested restricted shares outstanding as of December 31, 2017	853,126	$ 0.0001	4,468,753	$ 0.0001

The aggregate fair value of restricted shares that vested during the years ended December 31, 2016 and 2017 was $2,348 and $3,973, respectively.

Share-Based Compensation

Share-based compensation expense was classified in the consolidated statements of operations and comprehensive loss as follows:

	Year Ended December 31,	
	2016	2017
Research and development expenses	$ 59	$ 324
General and administrative expenses	309	573
	$ 368	$ 897

As of December 31, 2017, total unrecognized compensation cost related to the unvested share-based awards was $4,280, which is expected to be recognized over a weighted average period of 3.22 years.

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

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

license to certain background patent rights related 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 tiered royalties of high single-digit to low double-digit percentages on annual net sales of licensed products.

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.

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, the Company recorded research and development expense in connection with the Biogen Agreement of $12,100 and $4,169, 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.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

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. 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, the Company determined that the payment related to the milestone was not probable and, therefore, no amount was recorded in the Company's consolidated statement of operations and comprehensive loss during the year ended December 31, 2017. 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, the Company recorded research and development expense of $1,500 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. During the Option Period, the Company may conduct research and pre-clinical work to assess the viability of the asset.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

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.

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.

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 ARCALYST 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, the Company recognized research and development expense of $208 related to the purchase of drug materials under this agreement. As of December 31, 2017, the Company has non-cancelable purchase commitments under the clinical supply agreement (see Note 12).

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 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 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 an ARCALYST 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, the Company recorded research and development expense of $5,208 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 of low double-digit percentages based on annual net sales

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

9. License and Acquisition Agreements (Continued)

of products licensed under the agreement. 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.

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.

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.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

10. Income Taxes (Continued)

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)

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	0.0%	0.0%
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	—%	—%

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 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.

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)

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

10. Income Taxes (Continued)

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.

Basic and diluted net loss per share attributable to common shareholders was calculated as follows:

	Year Ended December 31,	
	2016	2017
Numerator:		
Net loss attributable to common shareholders	$ (23,973)	$ (64,873)
Denominator:		
Weighted average common shares outstanding — basic and diluted	715,045	4,944,889
Net loss per share attributable to common shareholders — basic and diluted	$ (33.53)	$ (13.12)

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

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)

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,	
	2016	2017
Options to purchase common shares	4,327,246	8,533,432
Unvested restricted shares	8,246,880	5,321,879
Convertible preferred shares (as converted to common shares)	46,800,044	62,531,219
	59,374,170	76,386,530

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 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 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 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.

Unaudited pro forma basic and diluted net loss per share attributable to common shareholders was calculated as follows:

	Year Ended December 31, 2017 (unaudited)
Numerator:	
Net loss attributable to common shareholders	$ (64,873)
Denominator:	
Weighted average common shares outstanding — basic and diluted	4,944,889
Pro forma adjustment to reflect assumed automatic conversion of Preferred Shares upon the closing of the proposed initial public offering	59,643,579
Pro forma weighted average common shares outstanding — basic and diluted	64,588,468
Pro forma net loss per share attributable to common shareholders — basic and diluted	$ (1.00)

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.

The Company recognizes rent expense on a straight-line basis over the respective lease period. The Company recorded rent expense of $286 and $402 during the years ended December 31, 2016 and 2017, 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, the Company entered into agreements with several contract manufacturing organizations to provide pre-clinical and clinical trial materials. As of December 31, 2017, the Company had non-cancelable purchase commitments under these agreements totaling $7,766 which are all due during the year ending December 31, 2018.

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.

KINIKSA PHARMACEUTICALS, LTD.

NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)

(Amounts in thousands, except share and per share amounts)

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, the Company contributed $143 and $264, 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.

Sale of Series C Preferred Shares

In February 2018, the Company issued and sold 34,932,049 Series C preferred shares at an issuance price of $5.7254 per share for gross proceeds of $200,000.

The rights and preferences of the Series C preferred shares are substantially similar to the Company's Preferred Shares. 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, each initially equal to $5.7254, in effect at the time of conversion. In the event of a mandatory conversion of Series C preferred shares as a result of a Qualified IPO, holders of Series C preferred shares may elect to receive Class A1 common shares in lieu of Class A common shares.

In February 2018, in connection with the Company's sale of Series C preferred shares, the Company amended and restated its Amended Bye-Laws to increase the total number of authorized shares of all classes of capital stock to 122,263,000 shares, consisting of 97,463,268 preferred shares, 15,049,615 Class A common shares and 9,750,005 Class B common shares.

Class A Common Shares

Class A Common Shares

Goldman, Sachs & Co. LLC
J.P. Morgan
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	$ *
FINRA filing fee	*
The Nasdaq Global Market initial listing fee	*
Accountants' fees and expenses	*
Legal fees and expenses	*
Blue Sky fees and expenses	*
Transfer Agent's fees and expenses	*
Printing and engraving expenses	*
Miscellaneous	*
Total expenses	$ *

* To be provided by amendment

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 1,950,001 shares of Class A common shares to our employees and a consultant at a price per share of $.0001 for aggregate gross consideration of $195.

In October 2015, we issued 9,750,005 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 21,937,521 Series A preferred shares to investors at a price per share of $1.7094 for aggregate gross consideration of $37.5 million.

In September 2016, we issued and sold 24,862,523 Series A preferred shares to investors at a price per share of $1.7094 for aggregate gross consideration of $42.5 million.

In March 2017, we issued and sold an aggregate of 15,731,175 Series B preferred shares to investors at a price per share of $2.5427 for aggregate gross consideration of $40.0 million.

In February 2018, we issued and sold an aggregate of 34,932,049 Series C preferred shares to investors at a price per $5.7254 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 17,241 Class A common shares pursuant to the exercise of share options at an exercise price of $0.58 for total aggregate proceeds to the registrant of $9,999.78.

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 upon 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, Bermuda
10.1	2015 Equity Incentive Plan, as amended, and form of option agreement thereunder
10.2*	2018 Incentive Award Plan and form of award agreements thereunder
10.3*	Form of Indemnification Agreement for Directors and Officers
10.4*	Amended and Restated Employment Agreement, dated as of June 29, 2017, by and between Kiniksa Pharmaceuticals Corp. and Sanjiv K. Patel
10.5*	Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Stephen F. Mahoney
10.6*	Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and John F. Paolini
10.7*	Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Chris Heberlig
10.8*	Employment Agreement by and between Kiniksa Pharmaceuticals Corp. and Thomas W. Beetham
10.9*	Lease Agreement for Kiniksa Pharmaceuticals Corp.
10.10†	Asset Purchase Agreement, dated September 7, 2016, by and between the Registrant and Biogen MA Inc., as amended
10.11†	License Agreement, dated September 25, 2017, by and between the Registrant and Regeneron Pharmaceuticals, Inc.
10.12†	License Agreement, dated as of December 21, 2017, by and between the Registrant and MedImmune, Limited
10.13	Clinical Supply Agreement, dated as of September 27, 2017, by and between the Registrant and Regeneron Pharmaceuticals, Inc.
21.1	Subsidiaries of the Registrant
23.1*	Consent of PricewaterhouseCoopers LLP, independent registered public accounting firm
23.2*	Consent of Conyers Dill & Pearman, Bermuda (included in Exhibit 5.1)

Exhibit Number	Description of Exhibit
24.1*	Power of Attorney (included in the signature pages to this Registration Statement)

* To be filed by amendment.

† 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 day of 2018.

KINIKSA PHARMACEUTICALS, LTD.

By: _____

Sanj K. Patel
Chief Executive Officer and Chairman of the Board of Directors

SIGNATURES

We, the undersigned officers and directors of Kiniksa Pharmaceuticals, Ltd., hereby severally constitute and appoint Sanj K. Patel and Chris Heberlig, and each of them singly (with full power to each of them to act alone), our true and lawful attorneys-in-fact and agents, with full power of substitution and resubstitution in each of them for him and in his name, place and stead, and in any and all capacities, to sign any and all amendments (including post-effective amendments) to this registration statement (or any other registration statement for the same offering that is to be effective upon filing pursuant to Rule 462(b) under the Securities Act of 1933), and to file the same, with all exhibits thereto and other documents in connection therewith, with the Securities and Exchange Commission, granting unto said attorneys-in-fact and agents, and each of them, full power and authority to do and perform each and every act and thing requisite or necessary to be done in and about the premises, as full to all intents and purposes as he might or could do in person, hereby ratifying and confirming all that said attorneys-in-fact and agents or any of them, or their or his substitute or substitutes, may lawfully do or cause to be done by virtue hereof.

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
Sanj K. Patel	Chief Executive Officer and Chairman of the Board of Directors (principal executive officer)	, 2018
Chris Heberlig	Chief Financial Officer (principal financial and accounting officer)	, 2018
Felix Baker, Ph.D.	Director	, 2018
Stephen R. Biggar, M.D., Ph.D.	Director	, 2018
Thomas Malley	Director	, 2018
Tracey McCain	Director	, 2018
Kimberly Popovits	Director	, 2018
Barry D. Quart, Pharm.D.	Director	, 2018

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 day of 2018.

KINIKSA PHARMACEUTICALS, CORP.

By: _____

Sanj K. Patel
Chief Executive Officer

Exhibit 3.1

FORM NO. 2

BERMUDA

THE COMPANIES ACT 1981

MEMORANDUM OF ASSOCIATION OF COMPANY LIMITED BY SHARES

(Section 7(1) and (2))

MEMORANDUM OF ASSOCIATION OF

Kiniksa Pharmaceuticals, Ltd.

(hereinafter referred to as "the Company")

1. The liability of the members of the Company is limited to the amount (if any) for the time being unpaid on the shares respectively held by them.

2. We, the undersigned, namely,

NAME	ADDRESS	BERMUDIAN STATUS (Yes/No)	NATIONALITY	NUMBER OF SHARES SUBSCRIBED
Graham B. R. Collis	Clarendon House 2 Church Street Hamilton HM 11 Bermuda	Yes	British	One
Jason Piney	"	No	British	One
David W. J. Astwood	"	Yes	British	One

do hereby respectively agree to take such number of shares of the Company as may be allotted to us respectively by the provisional directors of the Company, not exceeding the number of shares for which we have respectively subscribed, and to satisfy such calls as may be made by the directors, provisional directors or promoters of the Company in respect of the shares allotted to us respectively.

3. The Company is to be an **exempted** company as defined by the Companies Act 1981 (the "Act").

4. The Company, with the consent of the Minister of Finance, has power to hold land situate in Bermuda not exceeding ___ in all, including the following parcels:- **N/A**

5. The authorised share capital of the Company is **US$12,000.00** divided into shares of **US$0.0001** each.

6. The objects for which the Company is formed and incorporated are unrestricted.

7. The following are provisions regarding the powers of the Company –

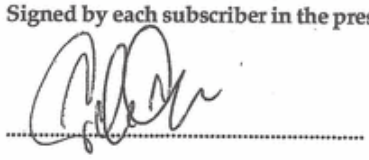
Subject to paragraph 6, the Company may do all such things as are incidental or conducive to the attainment of its objects and shall have the capacity, rights, powers and privileges of a natural person, and –

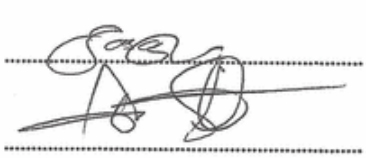
(i) pursuant to Section 42 of the Act, the Company shall have the power to issue preference shares which are, at the option of the holder, liable to be redeemed;

(ii) pursuant to Section 42A of the Act, the Company shall have the power to purchase its own shares for cancellation; and

(iii) pursuant to Section 42B of the Act, the Company shall have the power to acquire its own shares to be held as treasury shares.

Signed by each subscriber in the presence of at least one witness attesting the signature thereof





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(Subscribers)

(Witnesses)

SUBSCRIBED this 21st day of July, 2015.

Exhibit 3.2

AMENDED AND RESTATED BYE-LAWS

OF

KINIKSA PHARMACEUTICALS, LTD.

TABLE OF CONTENTS

Kiniksa Pharmaceuticals, 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;
Alternate Director	an alternate director appointed in accordance with these Bye-laws;

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 and shall include an Alternate Director;
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;
shareholder	a Member; 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 word “corporation” means a corporation whether or not a company within the meaning of the Act; and

(g) 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 and any shares or class of shares may be issued with such preferred, deferred or other special rights or such restrictions, whether in regard to dividend, voting, return of capital, or otherwise as the Company may by resolution of the Members prescribe.

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 A' Common Shares," "Class B Common Shares," "Class B' Common Shares" and "Preferred Shares." The total number of shares which the Company is authorised to issue is 122,263,000 shares, each with a par value of US$0.0001 per share. At this time, 15,049,615 shares shall be designated as Class A Common Shares, no shares shall be designated as Class A' Common Shares, 9,750,005 shares shall be designated as Class B Common Shares, no shares shall be designated as

Class B' Common Shares and 97,463,268 shares shall be designated as Preferred Shares. The undesignated portion of the share capital may be designated to any of the foregoing stated classes of shares at any time in the future by the Board. The Class A Common Shares, Class A' Common Shares, Class B Common Shares and Class B' 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 A' Common Shares, Class B Common Shares and Class B' 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 votes represented by all of the Company's outstanding issued 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 A' Common Shares and Class B' 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 votes represented by all of the Company's outstanding issued 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 B 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 Shares that were so converted and the number of Class A 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 B' 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 B' 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 B' 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 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 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 Shares that were so converted and the number of Class B' Shares shall increase in the same amount.

C. CLASS A' COMMON SHARES

(1) General. The Company shall not issue any Class A' Common Shares other than immediately prior to (or anytime thereafter) the effectiveness of a registration statement filed by the Company under the Securities Exchange Act of 1934, as amended. The 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 C, 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 A Common Shares, Class B Common Shares and Class B' Common Shares.

(2) Voting. The holders of the outstanding Class A' Common Shares shall possess no voting power whatsoever, either general or specific. The number of authorised Class A' 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 votes represented by all of the Company's outstanding issued shares entitled to vote.

(3) Optional Conversion. Each Class A' 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 A' Common Shares shall be prohibited from converting such shares if, as a result of such conversion (or portion of such conversion thereof), the holder, together with its affiliates, would own more than 9.99% of the voting power of the issued and outstanding Common Shares, which percentage may be increased or such limitation waived at such holder's election upon sixty-one (61) days' written notice to the Company. Before any holder of Class A' Common Shares shall be entitled to convert any of such Class A' 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 A' 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 A' Common Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Class A' 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 A' 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 A' 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 A' Common Share pursuant to the foregoing provisions, the number of authorised Class A' Common Shares shall be diminished by the number of Class A' Shares that were so converted and the number of Class A Shares shall increase in the same amount.

D. CLASS B' COMMON SHARES

(1) General. The Company shall not issue any Class B' Common Shares other than immediately prior to (or anytime thereafter) the effectiveness of a registration statement filed by the Company under the Securities Exchange Act of 1934, as amended. The 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 D, 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 A' Common Shares and Class B Common Shares.

(2) Voting. The holders of the outstanding Class B' Common Shares shall possess no voting power whatsoever, either general or specific. The number of authorised Class B' 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 votes represented by all of the Company's outstanding issued shares entitled to vote.

(3) Optional Conversion.

(3.1) 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; provided, however, that the holder of such Class B' Common Shares shall be prohibited from converting such shares if, as a result of such conversion (or portion of such conversion thereof), the holder, together with its affiliates, would own more than 9.99% of the voting power of the issued and outstanding Common Shares, which percentage may be increased or such limitation waived at such holder's election upon sixty-one (61) days' 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 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 B' 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' Shares that were so converted and the number of Class A Shares shall increase in the same amount.

(3.2) Each Class B' 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 B' Common Shares shall be prohibited from converting such shares if, as a result of such conversion (or portion of such conversion thereof), the holder, together with its affiliates, would own more than 9.99% of the voting power of the issued and outstanding Common Shares, which percentage may be increased or such limitation waived at such holder's election upon sixty-one (61) days' 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 B 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 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 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 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' Shares that were so converted and the number of Class B Shares shall increase in the same amount.

E. PREFERRED SHARES

The 97,463,268 authorised shares of the Company that are designated as Preferred Shares pursuant to Bye-law 4.1 are hereby further designated as follows: (a) 46,800,044 as “Series A Preferred Shares”, (b) 15,731,175 as “Series B Preferred Shares” and (c) 34,932,049 as “Series C Preferred Shares”. The Preferred Shares shall have the

following rights, preferences, powers, privileges and restrictions, qualifications and limitations. Unless otherwise indicated, references to “sections” or “subsections” in this Part E refer to sections and subsections of this Part E.

(1) Dividends. The Company shall not declare, pay or set aside any dividends on any other class or series of shares of the Company (payable other than in fully paid bonus Class A Common Shares, Class A’ Common Shares, Class B Common Shares, Class B’ Common Shares or other securities and rights convertible into or entitling the holder thereof to receive, directly or indirectly, additional Class A Common Shares, Class A’ Common Shares, Class B Common Shares or Class B’ Common Shares, respectively) unless (in addition to the obtaining of any consents required elsewhere in these Bye- laws) the holders of the Preferred Shares then issued and outstanding shall first receive, or simultaneously receive, a dividend on each outstanding Preferred Share in an amount at least equal to (i) in the case of a dividend on any class of Common Shares or any class or series that is convertible into a class of Common Shares, that dividend per Preferred Share as would equal the product of (A) 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 such class of Common Shares and (B) the number of such class of Common Shares issuable upon conversion of a share of the applicable series of Preferred Shares, in each case calculated on the record date for determination of holders entitled to receive such dividend or (ii) in the case of a dividend on any class or series that is not convertible into a class of Common Shares, at a rate per Preferred Share determined by (A) dividing the amount of the dividend payable on each share of such class or series of shares by the original issuance price of such class or series (subject to appropriate adjustment in the event of any bonus share, share dividend, share split, combination or other similar recapitalization with respect to such class or series) and (B) multiplying such fraction by an amount equal to, in the case of Series A Preferred Shares, the Series A Original Issue Price (as defined below), in the case of Series B Preferred Shares, the Series B Original Issue Price (as defined below), and, in the case of Series C Preferred Shares, the Series C Original Issue Price (as defined below); provided that, if the Company declares, pays or sets aside, on the same date, a dividend on more than one class or series of shares of the Company, the dividend payable to the holders of any series of Preferred Shares pursuant to this Section (1) shall be calculated based upon the dividend on the class or series of shares that would result in the highest Preferred Share dividend. The “Series A Original Issue Price” shall mean US$1.7094 per share, subject to appropriate adjustment in the event of any share dividend, share split, combination or other similar recapitalization with respect to the Series A Preferred Shares. The “Series B Original Issue Price” shall mean US$2.5427 per share, subject to appropriate adjustment in the event of any share dividend, share split, combination or other similar recapitalization with respect to the Series B Preferred Shares. The “Series C Original Issue Price” shall mean US$5.7254 per share, subject to appropriate adjustment in the event of any share dividend, share

split, combination or other similar recapitalization with respect to the Series C Preferred Shares.

(2) Liquidation, Dissolution or Winding Up; Certain Mergers, Consolidations and Asset Sales

(2.1) Preferential Payments to Holders of Preferred Shares. In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company or Deemed Liquidation Event, 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 (A) in the case of Series A Preferred Shares, the greater of (i) one (1) times the Series A Original Issue Price, plus any dividends declared but unpaid thereon, and (ii) such amount per share as would have been payable had all Series A Preferred Shares been converted into Class B Common Shares pursuant to Section (4) immediately prior to such liquidation, dissolution, winding up or Deemed Liquidation Event (the amount payable pursuant to this sentence is hereinafter referred to as the “Series A Liquidation Amount”), (B) in the case of Series B Preferred Shares, the greater of (i) one (1) times the Series B Original Issue Price, plus any dividends declared but unpaid thereon, and (ii) such amount per share as would have been payable had all Series B Preferred Shares been converted into Class A Common Shares pursuant to Section (4) immediately prior to such liquidation, dissolution, winding up or Deemed Liquidation Event (the amount payable pursuant to this sentence is hereinafter referred to as the “Series B Liquidation Amount”), and (C) in the case of Series C Preferred Shares, the greater of (i) one (1) times the 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 Series C Preferred Shares been converted into Class A Common Shares pursuant to Section (4) immediately prior to such liquidation, dissolution, winding up or Deemed Liquidation Event (the amount payable pursuant to this sentence is hereinafter referred to as the “Series C Liquidation Amount”). 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 under this Subsection (2.1), 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.

(2.2) Payments to Holders of Common Shares. In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company or Deemed Liquidation Event, after the payment of all preferential amounts required to be paid to

the holders of Preferred Shares, 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.

(2.3) Deemed Liquidation Events

(a) Definition. Each of the following events shall be considered a "Deemed Liquidation Event," unless 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 (the "Requisite Holders"), elect otherwise by written notice sent to the Company at least ten (10) days prior to the effective date of any such event:

(i) any amalgamation, merger or consolidation in which:

(A) the Company is a constituent party; or

(B) a subsidiary of the Company is a constituent party and the Company issues equity shares pursuant to such amalgamation, merger or consolidation,

except any such amalgamation, merger or consolidation involving the Company or a subsidiary in which the shares of the Company outstanding immediately prior to such amalgamation, merger or consolidation continue to represent, or are converted into or exchanged for shares that represent, immediately following such amalgamation, merger or consolidation, a majority, by voting power, of the equity shares of (1) the surviving or resulting company or corporation or (2) if the surviving or resulting company or corporation is a wholly owned subsidiary of another company or corporation immediately following such amalgamation, merger or consolidation, the parent company or corporation of such surviving or resulting company or corporation; or

(ii) the sale, lease, transfer, exclusive license or other disposition, in a single transaction or series of related transactions, by the Company or any subsidiary of the Company of all or substantially all the assets of the Company and its subsidiaries taken as a whole or the sale or disposition (whether by merger, consolidation or otherwise) of one or more subsidiaries of the Company if substantially all of the assets of the Company and its subsidiaries taken as a whole are held by such subsidiary or subsidiaries, except where such sale, lease, transfer, exclusive license or other disposition is to a wholly owned subsidiary of the Company.

(2.4) Effecting a Deemed Liquidation Event

(a) The Company shall not have the power to effect a Deemed Liquidation Event referred to in Subsection (2.3)(a)(i)(A) unless the agreement or plan of amalgamation, merger or consolidation for such transaction (the "Merger Agreement") provides that the consideration payable to the shareholders of the Company shall be allocated among the shareholders of the Company in accordance with Subsections (2.1) and (2.2).

(b) In the event of a Deemed Liquidation Event referred to in Subsection (2.3)(a)(i)(B) or (2.3)(a)(ii), if the Company does not effect a liquidation or dissolution of the Company under the Act within ninety (90) days after such Deemed Liquidation Event, then (i) the Company shall send a written notice to each holder of Preferred Shares no later than the ninetieth (90th) day after the Deemed Liquidation Event advising such holders of their right (and the requirements to be met to secure such right) pursuant to the terms of the following clause (ii) to require the redemption of such Preferred Shares, and (ii) if the Requisite Holders so request in a written instrument delivered to the Company not later than one hundred twenty (120) days after such Deemed Liquidation Event, the Company shall use the consideration received by the Company for such Deemed Liquidation Event (net of any retained liabilities associated with the assets sold or technology licensed, as determined in good faith by the Board), together with any other assets of the Company available for distribution to its members, all to the extent permitted by the provisions of the Act governing redemptions of shares and distributions to members (the "Available Proceeds"), on the one hundred fiftieth (150th) day after such Deemed Liquidation Event (the "Liquidation Redemption Date"), to redeem all issued and outstanding Preferred Shares at a price per share equal to, in the case of the Series A Preferred Shares, the Series A Liquidation Amount, in the case of the Series B Preferred Shares, the Series B Liquidation Amount, and, in the case of the Series C Preferred Shares, the Series C Liquidation Amount. Notwithstanding the foregoing, in the event of a redemption pursuant to the preceding sentence, if the Available Proceeds are not sufficient to redeem all outstanding Preferred Shares, the Company shall ratably redeem each holder's Preferred Shares to the fullest extent of such Available Proceeds, and shall redeem the remaining shares as soon as it may lawfully do so under the provisions of the Act governing redemptions of shares and distributions to shareholders. Prior to the distribution or redemption provided for in this Subsection (2.4)(b), the Company shall not expend or dissipate the consideration received for such Deemed Liquidation Event, except to discharge expenses incurred in connection with such Deemed Liquidation Event or in the ordinary course of business.

(c) Written notice of the mandatory redemption described in Subsection (2.4)(b) (the "Liquidation Redemption Notice") shall be mailed,

postage prepaid, to each holder of record of Preferred Shares, at its post office address last shown on the records of the Company, or given by electronic communication in compliance with the provisions of the Act, not less than twenty (20) days prior to the Liquidation Redemption Date. Each Liquidation Redemption Notice shall state:

(i) the number of Preferred Shares held by the holder that the Company shall redeem on the Liquidation Redemption Date specified in the Liquidation Redemption Notice;

(ii) the Liquidation Redemption Date and the Series A Liquidation Amount, the Series B Liquidation Amount and/or the Series C Liquidation Amount, as applicable; and

(iii) that the holder is to surrender to the Company, in the manner and at the place designated, his, her or its certificate or certificates representing the Preferred Shares to be redeemed.

(d) On or before the applicable Liquidation Redemption Date, each holder of Preferred Shares to be redeemed on such Liquidation Redemption Date, unless such holder has exercised his, her or its right to convert such shares as provided in Section 4 hereof, shall surrender the certificate or certificates representing such shares to the Company, in the manner and at the place designated in the Liquidation Redemption Notice, and thereupon the Series A Liquidation Amount, the Series B Liquidation Amount or the Series C Liquidation Amount, as the case may be, for such shares shall be payable to the order of the person whose name appears on such certificate or certificates as the owner thereof, and the shares represented by each such surrendered certificate shall be cancelled and retired. In the event less than all of the Preferred Shares represented by a certificate are redeemed, the Company's Register of Members shall be updated to reflect the number of Preferred Shares that were redeemed and a new share certificate representing the unredeemed Preferred Shares shall promptly be issued to such holder.

(e) If the Liquidation Redemption Notice shall have been duly given, and if on the Liquidation Redemption Date all Series A Liquidation Amounts, Series B Liquidation Amounts or Series C Liquidation Amounts, as applicable, payable upon redemption of the applicable Preferred Shares to be redeemed on such Liquidation Redemption Date are paid or tendered for payment or deposited with an independent payment agent so as to be available therefor, then notwithstanding that the certificates evidencing any of the Preferred Shares so called for redemption shall not have been surrendered, the Company's Register of Members shall be updated to give effect to such redemption of Preferred Shares, dividends with respect to such Preferred

Shares shall cease to accrue after such Liquidation Redemption Date and all rights with respect to such shares shall forthwith after the Liquidation Redemption Date terminate, except only the right of the holders to receive the Series A Liquidation Amount, Series B Liquidation Amount or Series C Liquidation Amount, as the case may be, per applicable Preferred Share without interest upon surrender of their certificate or certificates therefor.

(f) Any Preferred Shares which are redeemed or otherwise acquired by the Company or any of its subsidiaries shall be automatically and immediately cancelled and shall not be reissued, sold or transferred and shall thereafter form part of the Company's authorised, unissued and undesignated share capital. The Company may not exercise any voting or other rights granted to the holders of Preferred Shares following redemption.

(2.5) Amount Deemed Paid or Distributed. The amount deemed paid or distributed to the holders of equity shares of the Company upon any such amalgamation, merger, consolidation, sale, transfer, exclusive license, other disposition or redemption shall be the cash or the value of the property, rights or securities paid or distributed to such holders by the Company or the acquiring person, firm or other entity. The value of such property, rights or securities shall be determined in good faith by the Board.

(3) Voting

(3.1) General. On any matter presented to the shareholders of the Company for their action or consideration at any meeting of shareholders of the Company (or by written consent of shareholders in lieu of such meeting), (A) each holder of issued and outstanding Series A Preferred Shares shall be entitled to cast the number of votes equal to the product of (x) the number of whole Class B Common Shares into which the Series A Preferred Shares held by such holder are convertible as of the record date for determining shareholders entitled to vote on such matter and (y) the number of votes that the holder of a Class B Common Share is entitled to vote with respect to such Class B Common Share as of the record date of determining shareholders entitled to vote on such matter, (B) each holder of issued and outstanding Series B Preferred Shares shall be entitled to cast the number of votes equal to the product of (x) the number of whole Class A Common Shares into which the Series B Preferred Shares held by such holder are convertible as of the record date for determining shareholders entitled to vote on such matter and (y) the number of votes that the holder of a Class A Common Share is entitled to vote with respect to such Class A Common Share as of the record date of determining shareholders entitled to vote on such matter, and (C) each holder of issued and outstanding Series C Preferred Shares shall be entitled to cast the number of votes equal to the product of (x) the number of whole Class A Common Shares into which the Series C Preferred Shares held by such holder are convertible as of the record date for determining shareholders entitled to vote on such matter and (y)

the number of votes that the holder of a Class A Common Share is entitled to vote with respect to such Class A Common Share as of the record date of determining shareholders entitled to vote on such matter. Except as provided by law or by the other provisions of these Bye-laws, holders of Preferred Shares shall vote together with the holders of Class A Common Shares and Class B Common Shares as a single class.

(3.2) Election of Directors. The holders of record of the Preferred Shares, voting together as a single class as if all Series A Preferred Shares converted into Class B Common Shares and all Series B Preferred Shares and Series C Preferred Shares converted into Class A Common Shares, shall be entitled to elect two (2) directors of the Company (the "Preferred Directors") and the holders of record of the Class A Common Shares and Class B Common Shares, voting together as a single class, shall be entitled to elect one (1) director of the Company. Any director elected as provided in the preceding sentence may be removed without cause by, and only by, the affirmative vote of the holders of the class or series of shares entitled to elect such director or directors, given either at a special meeting of such shareholders duly called for that purpose or pursuant to a written consent of such shareholders. If the holders of Preferred Shares, on the one hand, and/or Class A Common Shares and Class B Common Shares, on the other hand, fail to elect a sufficient number of directors to fill all directorships for which they are entitled to elect directors pursuant to the first sentence of this Subsection (3.2), then any directorship

not so filled shall remain vacant until such time as the holders of Preferred Shares and/or Class A Common Shares and Class B Common Shares, as the case may be, elect a person to fill such directorship by vote or written consent in lieu of a meeting; and no such directorship may be filled by shareholders of the Company other than by the shareholders of the Company that are entitled to elect a person to fill such directorship, voting exclusively and as a separate class as set forth in the first sentence of this Subsection (3.2). The holders of record of the Class A Common Shares and Class B Common Shares and of any other class or series of voting shares (including each series of Preferred Shares), exclusively and voting together as a single class, shall be entitled to elect the balance of the total number of directors of the Company. At any meeting held for the purpose of electing a director, the presence in person or by proxy of the holders of a majority of the outstanding shares of the class or series entitled to elect such director shall constitute a quorum for the purpose of electing such director. Except as otherwise provided in this Subsection (3.2), a vacancy in any directorship filled by the holders of any class or series shall be filled only by vote or written consent in lieu of a meeting of the holders of such class or series or by any remaining director or directors elected by the holders of the same class or series pursuant to this Subsection (3.2). The rights of the holders of the Preferred Shares and the rights of the holders of the Class A Common Shares and Class B Common Shares under the first sentence of this Subsection (3.2) shall terminate on the first date following the Series C Original Issue Date (as defined below) on which there are issued and outstanding less than 29,239,129 Preferred Shares (subject to

appropriate adjustment in the event of any share dividend, share split, combination, or other similar recapitalization with respect to the Preferred Shares).

(3.3) Preferred Shares Protective Provisions. At any time when at least 4,873,188 Preferred Shares (subject to appropriate adjustment in the event of any bonus issue, share dividend, share split, combination or other similar recapitalization with respect to the Preferred Shares) are issued and outstanding, the Company shall not, either directly or indirectly by amendment, amalgamation, merger, consolidation or otherwise, do any of the following without (in addition to any other vote required by law or these Bye-laws) the written consent or affirmative vote of the Requisite Holders, given in writing or by vote at a meeting:

(a) liquidate, dissolve or wind-up the business and affairs of the Company, effect any amalgamation, merger or consolidation or any other Deemed Liquidation Event, or consent to any of the foregoing;

(b) amend, alter or repeal any provision of the Memorandum of Association or these Bye-laws;

(c) (i) create, or authorise the creation of, or issue any additional class or series of equity shares unless the same ranks junior to the Preferred Shares with respect to the distribution of assets on the liquidation, dissolution or winding up of the Company or the payment of dividends, (ii) increase the authorised number of Preferred Shares or (iii) increase the authorised number of any additional class or series of shares unless the same ranks junior to the Preferred Shares with respect to the distribution of assets on the liquidation, dissolution or winding up of the Company or the payment of dividends;

(d) (i) reclassify, alter or amend any existing security of the Company that is pari passu with the Preferred Shares in respect of the distribution of assets on the liquidation, dissolution or winding up of the Company or the payment of dividends, if such reclassification, alteration or amendment would render such other security senior to the Preferred Shares in respect of any such right, preference, or privilege or (ii) reclassify, alter or amend any existing security of the Company that is junior to the Preferred Shares in respect of the distribution of assets on the liquidation, dissolution or winding up of the Company or the payment of dividends, if such reclassification, alteration or amendment would render such other security senior to or pari passu with the Preferred Shares in respect of any such right, preference or privilege;

(e) purchase or redeem (or permit any subsidiary to purchase or redeem) or pay or declare any dividend or make any distribution on, any shares of the Company other than (i) redemptions of or dividends or

distributions on the Preferred Shares as expressly authorised herein and (ii) repurchases of shares from former employees, officers, directors, consultants or other persons who performed services for the Company or any subsidiary in connection with the cessation of such employment or service at the lower of the original purchase price or the then-current fair market value thereof or (iv) as approved by the Board, including the approval of each of the Preferred Directors;

(f) create, or authorise the creation of, or issue, or authorise the issuance of any debt security, or permit any subsidiary to take any such action with respect to any debt security unless such debt security has received the prior approval of the Board, including the approval of each of the Preferred Directors;

(g) create, or hold capital stock in, any subsidiary that is not wholly owned (either directly or through one or more other subsidiaries) by the Company, or sell, transfer or otherwise dispose of any capital stock of any direct or indirect subsidiary of the Company, or permit any direct or indirect subsidiary to sell, lease, transfer, exclusively license or otherwise dispose (in a single transaction or series of related transactions) of all or substantially all of the assets of such subsidiary; or

(h) increase or decrease the authorised number of directors constituting the Board.

(3.4) Series A Preferred Shares Protective Provisions. At any time when at least 2,340,002 Series A Preferred Shares (subject to appropriate adjustment in the event of any bonus issue, share dividend, share split, combination or other similar recapitalization with respect to the Series A Preferred Shares) are issued and outstanding, the Company shall not, either directly or indirectly by amendment, amalgamation, merger, consolidation or otherwise, do any of the following without (in addition to any other vote required by law or these Bye-laws) the written consent or affirmative vote of the holders of a majority of the Series A Preferred Shares, voting exclusively as a class, given in writing or by vote at a meeting:

(a) amend, alter or repeal any provision of the Memorandum of Association or these Bye-laws so as to change the preferences, rights, privileges or powers of the Series A Preferred Shares adversely and in a manner disproportionate to the effect of the same on any other series of Preferred Shares; or

(b) increase or decrease the authorised number of Series A Preferred Shares.

(3.5) Series B Preferred Shares Protective Provisions. At any time when at least 786,566 Series B Preferred Shares (subject to appropriate adjustment in the event of any bonus issue, share dividend, share split, combination or other similar recapitalization with respect to the Series B Preferred Shares) are issued and outstanding, the Company shall not, either directly or indirectly by amendment, amalgamation, merger, consolidation or otherwise, do any of the following without (in addition to any other vote required by law or these Bye-laws) the written consent or affirmative vote of the holders of a majority of the Series B Preferred Shares, voting exclusively as a class, given in writing or by vote at a meeting:

(a) amend, alter or repeal any provision of the Memorandum of Association or these Bye-laws so as to change the preferences, rights, privileges or powers of the Series B Preferred Shares adversely and in a manner disproportionate to the effect of the same on any other series of Preferred Shares; or

(b) increase or decrease the authorised number of Series B Preferred Shares.

(3.6) Series C Preferred Shares Protective Provisions. At any time when at least 1,746,632 Series C Preferred Shares (subject to appropriate adjustment in the event of any bonus issue, share dividend, share split, combination or other similar recapitalization with respect to the Series C Preferred Shares) are issued and outstanding, the Company shall not, either directly or indirectly by amendment, amalgamation, merger, consolidation or otherwise, do any of the following without (in addition to any other vote required by law or these Bye-laws) the written consent or affirmative vote of the holders of a majority of the Series C Preferred Shares, voting exclusively as a class, given in writing or by vote at a meeting:

(a) amend, alter or repeal any provision of the Memorandum of Association or these Bye-laws so as to change the preferences, rights, privileges or powers of the Series C Preferred Shares adversely and in a manner disproportionate to the effect of the same on any other series of Preferred Shares; or

(b) increase or decrease the authorised number of Series C Preferred Shares.

(4) Optional Conversion

The holders of the Preferred Shares shall have conversion rights as follows (the “Conversion Rights”):

(4.1) Right to Convert

(a) Conversion Ratio. Each Series A Preferred Share shall be convertible, at the option of the holder thereof, 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 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 thereof, 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 thereof, 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 Conversion Price” shall initially be equal to US$1.7094, the “Series B Conversion Price” shall initially be equal to US$2.5427 and the “Series C Conversion Price” shall initially be equal to US$5.7254. The Series A Conversion Price, Series B Conversion Price and Series C Conversion Price are each referred to herein as a “Conversion Price.” Such initial Series A Conversion Price, Series B Conversion Price and Series C Conversion Price, and the rate at which shares of each series of Preferred Shares may be converted into Common Shares, shall be subject to adjustment as provided below.

(b) Termination of Conversion Rights. In the event of a notice of redemption of any Preferred Shares pursuant to Subsection (2.4)(c), the Conversion Rights of the shares designated for redemption shall terminate at the close of business on the last full day preceding the date fixed for redemption, unless the redemption price is not fully paid on such redemption date, in which case the Conversion Rights for such shares shall continue until such price is paid in full. In the event of a liquidation, dissolution or winding up of the Company or a Deemed Liquidation Event, the Conversion Rights shall terminate at the close of business on the last full day preceding the date fixed for the payment of any such amounts distributable on such event to the holders of Preferred Shares.

(4.2) Fractional Shares. No fractional Common Shares shall be issued upon conversion of any Preferred Shares. In lieu of any fractional shares to which the holder would otherwise be entitled, the Company shall pay cash equal to such fraction

multiplied by the fair market value of a share of the applicable class of Common Share as determined in good faith by the Board. Whether or not, and to the extent that, fractional shares would be issuable upon such conversion shall be determined on the basis of the total number of Preferred Shares the holder is at the time converting into each class of Common Shares and the aggregate number of such class(es) of Common Shares issuable upon such conversion.

(4.3) Mechanics of Conversion.

(a) Notice of Conversion. In order for a holder of Preferred Shares to voluntarily convert such Preferred Shares into a class of Common Shares, such holder shall (a) provide written notice to the Company's transfer agent at the office of the transfer agent for the Preferred Shares (or at the principal office of the Company if the Company serves as its own transfer agent) that such holder elects to convert all or any number of such holder's Preferred Shares and, if applicable, any event on which such conversion is contingent and (b), if such holder's shares are certificated, surrender the certificate or certificates for such Preferred Shares (or, if such registered holder alleges that such certificate has been lost, stolen or destroyed, a lost certificate affidavit and agreement reasonably acceptable to the Company to indemnify the Company against any claim that may be made against the Company on account of the alleged loss, theft or destruction of such certificate), at the office of the transfer agent for the Preferred Shares (or at the principal office of the Company if the Company serves as its own transfer agent). Such notice shall state such holder's name or the names of the nominees in which such holder wishes the applicable Common Shares to be issued. If required by the Company, any certificates surrendered for conversion shall be endorsed or accompanied by a written instrument or instruments of transfer, in form satisfactory to the Company, duly executed by the registered holder or his, her or its attorney duly authorised in writing. The close of business on the date of receipt by the transfer agent (or by the Company if the Company serves as its own transfer agent) of such notice and, if applicable, certificates (or lost certificate affidavit and agreement) shall be the time of conversion (the "Conversion Time"), and the Common Shares issuable upon conversion of the specified Preferred Shares shall be deemed to be issued and outstanding of record as of such date. The Company shall, as soon as practicable after the Conversion Time, (i) convert such Preferred Shares by updating its Register of Members and upon such action, to the extent permitted by applicable law, each affected Preferred Share shall be converted into and shall become (in such manner as is permitted by Bermuda law) the requisite number of fully paid and non-assessable Class A Common Shares, Class A' Common Shares, Class B Common Shares or Class B' Common Shares, as the case may be; (ii) issue and deliver to such holder of Preferred Shares, or to his, her or its nominees, a certificate or certificates for the

number of full Common Shares issuable upon such conversion in accordance with the provisions hereof and a certificate for the number (if any) of the Preferred Shares represented by the surrendered certificate that were not converted into Common Shares, (iii) pay in cash such amount as provided in Subsection (4.2) in lieu of any fraction of a Common Share otherwise issuable upon such conversion and (iv) pay all declared but unpaid dividends on the Preferred Shares converted.

(b) Reservation of Shares. The Company shall at all times when the Preferred Shares shall be outstanding, reserve and keep available out of its authorised but unissued shares, for the purpose of effecting the conversion of the Preferred Shares (if so required by applicable law), such number of its duly authorised Class A Common Shares, Class A' Common Shares, Class B Common Shares and Class B' Common Shares as shall from time to time be sufficient to effect the conversion of all outstanding Preferred Shares; and if at any time the number of authorised but unissued Class A Common Shares, Class A' Common Shares, Class B Common Shares or Class B' Common Shares shall not be sufficient to effect the conversion of all then outstanding Preferred Shares, the Company shall take such corporate action as may be necessary to increase its authorised but unissued Common Shares (or applicable class(es) thereof) to such number of shares as shall be sufficient for such purposes, including, without limitation, engaging in best efforts to obtain the requisite shareholder approval of any necessary amendment to the Memorandum of Association. Before taking any action which would cause an adjustment reducing a Conversion Price below the then par value of the applicable class of Common Shares issuable upon conversion of such series of Preferred Shares, the Company will take any corporate action which may, in the opinion of its counsel, be necessary in order that the Company may validly and legally issue fully paid and non-assessable Class A Common Shares, Class A' Common Shares, Class B Common Shares or Class B' Common Shares Shares, as the case may be, at such adjusted Conversion Price.

(c) Effect of Conversion. All Preferred Shares which shall have been surrendered for conversion as herein provided shall no longer be deemed to be issued and outstanding as Preferred Shares and all rights with respect to such shares shall immediately cease and terminate at the Conversion Time, except only the right of the holders thereof to receive the applicable class of Common Shares on conversion thereof, to receive payment in lieu of any fraction of a share otherwise issuable upon such conversion as provided in Subsection (4.2) and to receive payment of any dividends declared but unpaid thereon. It is intended that all Preferred Shares so converted shall convert into and shall become shares of the applicable class of Common Shares and shall cease to exist as Preferred Shares. Upon the conversion of any Preferred Share pursuant to

the foregoing provisions, the number of authorised Series A Preferred Shares, Series B Preferred Shares or Series C Preferred Shares, as applicable, shall be diminished by the number of such series of Preferred Shares that were so converted and the number of shares of the applicable class(es) of Common Shares shall increase in the same amount.

(d) No Further Adjustment. Upon any such conversion, no adjustment to the applicable Conversion Price(s) shall be made for any declared but unpaid dividends on the applicable series of Preferred Shares surrendered for conversion or on the applicable class(es) of Common Shares delivered upon conversion.

(e) Taxes. The Company shall pay any and all issue and other similar taxes that may be payable in respect of any issuance or delivery of Common Shares upon conversion of Preferred Shares pursuant to this Section (4). The Company shall not, however, be required to pay any tax which may be payable in respect of any transfer involved in the issuance and delivery of Common Shares in a name other than that in which the Preferred Shares so converted were registered, and no such issuance or delivery shall be made unless and until the person or entity requesting such issuance has paid to the Company the amount of any such tax or has established, to the satisfaction of the Company, that such tax has been paid.

(4.4) Adjustments to Conversion Prices for Diluting Issues

(a) Special Definitions. For purposes of this Part E, the following definitions shall apply:

(i) "Option" shall mean rights, options or warrants to subscribe for, purchase or otherwise acquire Common Shares or Convertible Securities.

(ii) "Series C Original Issue Date" shall mean the date on which the first Series C Preferred Share was issued.

(iii) "Convertible Securities" shall mean any evidences of indebtedness, shares or other securities directly or indirectly convertible into or exchangeable for Common Shares, but excluding Options.

(iv) "Additional Shares" shall mean all Common Shares issued (or, pursuant to Subsection (4.4)(c) below, deemed to be issued) by the Company after the Series C Original Issue Date, other than (1) the following Common Shares and (2) Common Shares deemed issued

pursuant to the following Options and Convertible Securities (clauses (1) and (2), collectively, "Exempted Securities"):

(A) Common Shares, Options or Convertible Securities issued as a dividend or distribution on Preferred Shares;

(B) Common Shares, Options or Convertible Securities issued by reason of a dividend, share split, split-up or other distribution on Common Shares that is covered by Subsection (4.5), (4.6), (4.7) or (4.8);

(C) Common Shares or Options issued to employees or directors of, or consultants or advisors to, the Company or any of its subsidiaries pursuant to a plan, agreement or arrangement approved by the Board, including each of the Preferred Directors;

(D) Common Shares or Convertible Securities actually issued upon the exercise of Options or Common Shares actually issued upon the conversion or exchange of Convertible Securities, in each case provided such issuance is pursuant to the terms of such Option or Convertible Security;

(E) Common Shares Options or Convertible Securities issued to banks, equipment lessors or other financial institutions, or to real property lessors, pursuant to a debt financing, equipment leasing or real property leasing transaction approved by the Board, including each of the Preferred Directors;

(F) Common Shares, Options or Convertible Securities issued in connection with supplier, third-party service provider, sponsored research, collaboration, license, development, marketing or other similar agreements or strategic partnerships, in each case approved by the Board, including each of the Preferred Directors; or

(G) Common Shares, Options or Convertible Securities issued pursuant to the acquisition of another corporation by the Company by merger, purchase of substantially all of the assets or other reorganization or to a joint venture agreement, provided that such issuances are approved by the Board, including each of the Preferred Directors.

(b) No Adjustment of Conversion Prices. No adjustment in the Series A Conversion Price shall be made as the result of the issuance or deemed issuance of Additional Shares if the Company receives written notice from the holders of a majority of the then outstanding Series A Preferred Shares, voting exclusively as a class, agreeing that no such adjustment shall be made as the result of the issuance or deemed issuance of such Additional Shares. No adjustment in the Series B Conversion Price shall be made as the result of the issuance or deemed issuance of Additional Shares if the Company receives written notice from the holders of a majority of the then outstanding Series B Preferred Shares, voting exclusively as a class, agreeing that no such adjustment shall be made as the result of the issuance or deemed issuance of such Additional Shares. No adjustment in the Series C Conversion Price shall be made as the result of the issuance or deemed issuance of Additional Shares if the Company receives written notice from the holders of a majority of the then outstanding Series C Preferred Shares, voting exclusively as a class, agreeing that no such adjustment shall be made as the result of the issuance or deemed issuance of such Additional Shares.

(c) Deemed Issue of Additional Shares.

(i) If the Company at any time or from time to time after the Series C Original Issue Date shall issue any Options or Convertible Securities (excluding Options or Convertible Securities which are themselves Exempted Securities) or shall fix a record date for the determination of holders of any class of securities entitled to receive any such Options or Convertible Securities, then the maximum number of Common Shares (as set forth in the instrument relating thereto, assuming the satisfaction of any conditions to exercisability, convertibility or exchangeability but without regard to any provision contained therein for a subsequent adjustment of such number) issuable upon the exercise of such Options or, in the case of Convertible Securities and Options therefor, the conversion or exchange of such Convertible Securities, shall be deemed to be Additional Shares issued as of the time of such issue or, in case such a record date shall have been fixed, as of the close of business on such record date.

(ii) If the terms of any Option or Convertible Security, the issuance of which resulted in an adjustment to a Conversion Price pursuant to the terms of Subsection (4.4)(d), are revised as a result of an amendment to such terms or any other adjustment pursuant to the provisions of such Option or Convertible Security (but excluding automatic adjustments to such terms pursuant to anti-dilution or similar provisions of such Option or Convertible Security) to provide for either

(1) any increase or decrease in the number of Common Shares issuable upon the exercise, conversion and/or exchange of any such Option or Convertible Security or (2) any increase or decrease in the consideration payable to the Company upon such exercise, conversion and/or exchange, then, effective upon such increase or decrease becoming effective, the applicable Conversion Price computed upon the original issue of such Option or Convertible Security (or upon the occurrence of a record date with respect thereto) shall be readjusted to such Conversion Price as would have been obtained had such revised terms been in effect upon the original date of issuance of such Option or Convertible Security. Notwithstanding the foregoing, no readjustment pursuant to this Subsection (4.4)(c)(ii) shall have the effect of increasing a Conversion Price to an amount which exceeds the lower of (i) the applicable Conversion Price in effect immediately prior to the original adjustment made as a result of the issuance of such Option or Convertible Security, or (ii) the applicable Conversion Price that would have resulted from any issuances of Additional Shares (other than deemed issuances of Additional Shares as a result of the issuance of such Option or Convertible Security) between the original adjustment date and such readjustment date.

(iii) If the terms of any Option or Convertible Security (excluding Options or Convertible Securities which are themselves Exempted Securities), the issuance of which did not result in an adjustment to a Conversion Price pursuant to the terms of Subsection (4.4)(d) (either because the consideration per share (determined pursuant to Subsection (4.4)(e) of the Additional Shares subject thereto was equal to or greater than the applicable Conversion Price then in effect, or because such Option or Convertible Security was issued before the Series C Original Issue Date), are revised after the Series C Original Issue Date as a result of an amendment to such terms or any other adjustment pursuant to the provisions of such Option or Convertible Security (but excluding automatic adjustments to such terms pursuant to anti-dilution or similar provisions of such Option or Convertible Security) to provide for either (1) any increase in the number of Common Shares issuable upon the exercise, conversion or exchange of any such Option or Convertible Security or (2) any decrease in the consideration payable to the Company upon such exercise, conversion or exchange, then such Option or Convertible Security, as so amended or adjusted, and the Additional Shares subject thereto (determined in the manner provided in Subsection (4.4)(c)(i)) shall be deemed to have been issued effective upon such increase or decrease becoming effective.

(iv) Upon the expiration or termination of any unexercised Option or unconverted or unexchanged Convertible Security (or portion thereof) which resulted (either upon its original issuance or upon a revision of its terms) in an adjustment to a Conversion Price pursuant to the terms of Subsection (4.4)(d), the applicable Conversion Price shall be readjusted to such applicable Conversion Price as would have been obtained had such Option or Convertible Security (or portion thereof) never been issued.

(v) If the number of Common Shares issuable upon the exercise, conversion and/or exchange of any Option or Convertible Security, or the consideration payable to the Company upon such exercise, conversion and/or exchange, is calculable at the time such Option or Convertible Security is issued or amended but is subject to adjustment based upon subsequent events, any adjustment to a Conversion Price provided for in this Subsection (4.4)(c) shall be effected at the time of such issuance or amendment based on such number of shares or amount of consideration without regard to any provisions for subsequent adjustments (and any subsequent adjustments shall be treated as provided in subsections (ii) and (iii) of this Subsection (4.4)(c)). If the number of Common Shares issuable upon the exercise, conversion and/or exchange of any Option or Convertible Security, or the consideration payable to the Company upon such exercise, conversion and/or exchange, cannot be calculated at all at the time such Option or Convertible Security is issued or amended, any adjustment to a Conversion Price that would result under the terms of this Subsection (4.4)(c) at the time of such issuance or amendment shall instead be effected at the time such number of shares and/or amount of consideration is first calculable (even if subject to subsequent adjustments), assuming for purposes of calculating such adjustment to the applicable Conversion Price that such issuance or amendment took place at the time such calculation can first be made.

(d) Adjustment of Conversion Prices Upon Issuance of Additional Shares. In the event the Company shall at any time after the Series C Original Issue Date issue Additional Shares (including Additional Shares deemed to be issued pursuant to Subsection (4.4)(c)), without consideration or for a consideration per share less than a Conversion Price in effect immediately prior to such issue, then the applicable Conversion Price shall be reduced, concurrently with such issue, to a price (calculated to the nearest one-hundredth of a cent) determined in accordance with the following formula:

CP2 = CP1* (A + B) ÷ (A + C)

For purposes of the foregoing formula, the following definitions shall apply:

(i) "CP2" shall mean the applicable Conversion Price in effect immediately after such issue of Additional Shares;

(ii) "CP1" shall mean the applicable Conversion Price in effect immediately prior to such issue of Additional Shares;

(iii) "A" shall mean the number of Common Shares outstanding immediately prior to such issue of Additional Shares (treating for this purpose as outstanding all Common Shares issuable upon exercise of Options outstanding immediately prior to such issue or upon conversion or exchange of Convertible Securities (including the Preferred Shares) outstanding (assuming exercise of any outstanding Options therefor) immediately prior to such issue);

(iv) "B" shall mean the number of Common Shares that would have been issued if such Additional Shares had been issued at a price per share equal to CP1 (determined by dividing the aggregate consideration received by the Company in respect of such issue by CP1); and

(v) "C" shall mean the number of such Additional Shares issued in such transaction.

(e) Determination of Consideration. For purposes of this Subsection (4.4), the consideration received by the Company for the issue of any Additional Shares shall be computed as follows:

(i) Cash and Property: Such consideration shall:

(A) insofar as it consists of cash, be computed at the aggregate amount of cash received by the Company, excluding amounts paid or payable for accrued interest;

(B) insofar as it consists of property other than cash, be computed at the fair market value thereof at the time of such issue, as determined in good faith by the Board; and

(C) in the event Additional Shares are issued together with other shares or securities or other assets of the Company for consideration which covers both, be the proportion of such consideration so received, computed as provided in clauses (i) and (ii) above, as determined in good faith by the Board.

(ii) Options and Convertible Securities. The consideration per share received by the Company for Additional Shares deemed to have been issued pursuant to Subsection (4.4)(c), relating to Options and Convertible Securities, shall be determined by dividing:

(A) The total amount, if any, received or receivable by the Company as consideration for the issue of such Options or Convertible Securities, plus the minimum aggregate amount of additional consideration (as set forth in the instruments relating thereto, without regard to any provision contained therein for a subsequent adjustment of such consideration) payable to the Company upon the exercise of such Options or the conversion or exchange of such Convertible Securities, or in the case of Options for Convertible Securities, the exercise of such Options for Convertible Securities and the conversion or exchange of such Convertible Securities, by

(B) the maximum number Common Shares (as set forth in the instruments relating thereto, without regard to any provision contained therein for a subsequent adjustment of such number) issuable upon the exercise of such Options or the conversion or exchange of such Convertible Securities, or in the case of Options for Convertible Securities, the exercise of such Options for Convertible Securities and the conversion or exchange of such Convertible Securities.

(f) Multiple Closing Dates. In the event the Company shall issue on more than one date Additional Shares that are a part of one transaction or a series of related transactions and that would result in an adjustment to a Conversion Price pursuant to the terms of Subsection (4.4)(d), and such issuance dates occur within a period of no more than ninety (90) days from the first such issuance to the final such issuance, then, upon the final such issuance, the applicable Conversion Price shall be readjusted to give effect to all such issuances as if they occurred on the date of the first such issuance (and without giving effect to any additional adjustments as a result of any such subsequent issuances within such period).

(4.5) Adjustment for Share Splits and Combinations. If the Company shall at any time or from time to time after the Series C Original Issue Date effect a subdivision of the outstanding Common Shares, each Conversion Price in effect immediately before that subdivision shall be proportionately decreased so that the number of Common Shares issuable on conversion of each share of such series shall be increased in proportion to such increase in the aggregate number of Common Shares outstanding. If the Company shall at any time or from time to time after the Series C

Original Issue Date combine the outstanding Common Shares, each Conversion Price in effect immediately before the combination shall be proportionately increased so that the number of Common Shares issuable on conversion of each share of such series shall be decreased in proportion to such decrease in the aggregate number of Common Shares outstanding. Any adjustment under this subsection shall become effective at the close of business on the date the subdivision or combination becomes effective.

(4.6) Adjustment for Certain Dividends and Distributions. In the event the Company at any time or from time to time after the Series C Original Issue Date shall make or issue, or fix a record date for the determination of holders of Common Shares entitled to receive, a dividend or other distribution payable on the Common Shares in additional Common Shares, then and in each such event each Conversion Price in effect immediately before such event shall be decreased as of the time of such issuance or, in the event such a record date shall have been fixed, as of the close of business on such record date, by multiplying the applicable Conversion Price then in effect by a fraction:

(x) the numerator of which shall be the total number of Common Shares issued and outstanding immediately prior to the time of such issuance or the close of business on such record date, and

(y) the denominator of which shall be the total number of Common Shares issued and outstanding immediately prior to the time of such issuance or the close of business on such record date plus the number of Common Shares issuable in payment of such dividend or distribution.

Notwithstanding the foregoing: (a) if such record date shall have been fixed and such dividend is not fully paid or if such distribution is not fully made on the date fixed therefor, each Conversion Price shall be recomputed accordingly as of the close of business on such record date and thereafter such Conversion Prices shall be adjusted pursuant to this subsection as of the time of actual payment of such dividends or distributions; and (b) no such adjustment shall be made if the holders of the applicable series of Preferred Shares simultaneously receive a dividend or other distribution of the same class of Common Shares in a number equal to the number Common Shares as they would have received if all outstanding shares of such series of Preferred Shares had been converted into such class of Common Shares on the date of such event.

(4.7) Adjustments for Other Dividends and Distributions. In the event the Company at any time or from time to time after the Series C Original Issue Date shall make or issue, or fix a record date for the determination of holders of Common Shares entitled to receive, a dividend or other distribution payable in securities of the Company (other than a distribution of Common Shares in respect of Common Shares) or in other property and the provisions of Section (1) do not apply to such dividend or distribution, then and in each such event the holders of each series of Preferred Shares

shall receive, simultaneously with the distribution to the holders of Common Shares, a dividend or other distribution of such securities or other property in an amount equal to the amount of such securities or other property as they would have received if all outstanding shares of such series of Preferred Shares had been converted into Common Shares on the date of such event.

(4.8) Adjustment for Merger or Reorganization, etc. Subject to the provisions of Subsection (2), if there shall occur any reorganization, recapitalization, reclassification, consolidation or merger involving the Company in which the Common Shares (but not the Preferred Shares) are converted into or exchanged for securities, cash or other property (other than a transaction covered by Subsections (4.5), (4.6) or (4.7)), then, following any such reorganization, recapitalization, reclassification, consolidation or merger, each Preferred Share shall thereafter be convertible in lieu of the Common Shares into which it was convertible prior to such event into the kind and amount of securities, cash or other property which a holder of the number of Common Shares of the Company issuable upon conversion of one Series A Preferred Share, Series B Preferred Share or Series C Preferred Share, as applicable, immediately prior to such reorganization, recapitalization, reclassification, consolidation or merger would have been entitled to receive pursuant to such transaction; and, in such case, appropriate adjustment (as determined in good faith by the Board) shall be made in the application of the provisions in this Section (4) with respect to the rights and interests thereafter of the holders of the Preferred Shares, to the end that the provisions set forth in this Section (4) (including provisions with respect to changes in and other adjustments of the applicable Conversion Price) shall thereafter be applicable, as nearly as reasonably may be, in relation to any securities or other property thereafter deliverable upon the conversion of the Preferred Shares.

(4.9) Certificate as to Adjustments. Upon the occurrence of each adjustment or readjustment of a Conversion Price pursuant to this Section (4), the Company at its expense shall, as promptly as reasonably practicable but in any event not later than ten (10) days thereafter, compute such adjustment or readjustment in accordance with the terms hereof and furnish to each holder of the affected series of Preferred Shares a certificate setting forth such adjustment or readjustment (including the kind and amount of securities, cash or other property into which the affected series of Preferred Shares are convertible) and showing in detail the facts upon which such adjustment or readjustment is based. The Company shall, as promptly as reasonably practicable after the written request at any time of any holder of Preferred Shares (but in any event not later than ten (10) days thereafter), furnish or cause to be furnished to such holder a certificate setting forth (i) the applicable Conversion Price(s) then in effect, and (ii) the number of Common Shares and the amount, if any, of other securities, cash or property which then would be received upon the conversion of such series of Preferred Shares.

(4.10) Notice of Record Date. In the event:

(a) the Company shall take a record of the holders of any class of Common Shares (or other equity shares or securities at the time issuable upon conversion of any series of Preferred Shares) for the purpose of entitling or enabling them to

receive any dividend or other distribution, or to receive any right to subscribe for or purchase any shares of any class or any other securities, or to receive any other security; or

(b) of any capital reorganization of the Company, any reclassification of any class of Common Shares or any Deemed Liquidation Event; or

(c) of the voluntary or involuntary dissolution, liquidation or winding-up of the Company,

then, and in each such case, the Company will send or cause to be sent to the holders of Preferred Shares a notice specifying, as the case may be, (i) the record date for such dividend, distribution or right, and the amount and character of such dividend, distribution or right, or (ii) the effective date on which such reorganization, reclassification, consolidation, merger, transfer, dissolution, liquidation or winding-up is proposed to take place, and the time, if any is to be fixed, as of which the holders of record of the applicable class(es) of Common Shares (or such other equity shares or securities at the time issuable upon the conversion of the applicable series of Preferred Shares) shall be entitled to exchange their Common Shares (or such other equity shares or securities) for securities or other property deliverable upon such reorganization, reclassification, consolidation, merger, transfer, dissolution, liquidation or winding-up, and the amount per share and character of such exchange applicable to the Preferred Shares and the Common Shares. Such notice shall be sent at least ten (10) days prior to the record date or effective date for the event specified in such notice.

(5) Mandatory Conversion.

(5.1) Trigger Events. 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 $5.7254 per share (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 IPO resulting in at least $100 million of gross proceeds to the Company or (b) the date and time, or the occurrence of an event, specified by vote or written consent of the Requisite Holders (the time of such closing or the date and time specified or the time of the event specified in such vote or written consent is referred to herein as the "Mandatory Conversion Time"), then (x) 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, (y) all outstanding Series B Preferred Shares shall automatically be converted, in such manner as is permitted pursuant to Bermuda law, into Class A Common Shares, and (z) all outstanding Series

C Preferred Shares shall automatically be converted, in such manner as is permitted pursuant to Bermuda law, into Class A Common Shares, in each case at the then-effective conversion rate as calculated pursuant to Subsection (4.1)(a).

(5.2) Procedural Requirements. All holders of record of Preferred Shares shall be sent written notice of the Mandatory Conversion Time and the place designated for mandatory conversion of all such Preferred Shares pursuant to this Section (5). Such notice need not be sent in advance of the occurrence of the Mandatory Conversion Time. Upon receipt of such notice, each holder of Preferred Shares in certificated form shall surrender his, her or its certificate or certificates for all such shares (or, if such holder alleges that such certificate has been lost, stolen or destroyed, a lost certificate affidavit and agreement reasonably acceptable to the Company to indemnify the Company against any claim that may be made against the Company on account of the alleged loss, theft or destruction of such certificate) to the Company at the place designated in such notice. If so required by the Company, any certificates surrendered for conversion shall be endorsed or accompanied by written instrument or instruments of transfer, in form satisfactory to the Company, duly executed by the registered holder or by his, her or its attorney duly authorised in writing. All rights with respect to the Preferred Shares converted pursuant to Subsection (5.1), including the rights, if any, to receive notices and vote (other than as a holder of Common Shares), will terminate at the Mandatory Conversion Time (notwithstanding the failure of the holder or holders thereof to surrender any certificates at or prior to such time), except only the rights of the holders thereof, upon surrender of any certificate or certificates of such holders (or lost certificate affidavit and agreement) therefor, to receive the items provided for in the next sentence of this Subsection (5.2). As soon as practicable after the Mandatory Conversion Time and, if applicable, the surrender of any certificate or certificates (or lost certificate affidavit and agreement) for Preferred Shares, the Company shall (a) (1) issue and deliver to each holder of Series A Preferred Shares, or to his, her or its nominees, a certificate or certificates for the number of full Class B Common Shares issuable on conversion of such Series A Preferred Shares in accordance with the provisions hereof and (2) pay

cash as provided in Subsection (4.2) in lieu of any fraction of a Class B Common Share otherwise issuable upon such conversion and the payment of any declared but unpaid dividends on the Series A Preferred Shares so converted, (b) (1) issue and deliver to each holder of Series B Preferred Shares, or to his, her or its nominees, a certificate or certificates for the number of full Class A Common Shares issuable on conversion of such Series B Preferred Shares in accordance with the provisions hereof and (2) pay cash as provided in Subsection (4.2) in lieu of any fraction of a Class A Common Share otherwise issuable upon such conversion and the payment of any declared but unpaid dividends on the Series B Preferred Shares so converted and (c) (1) issue and deliver to each holder of Series C Preferred Shares, or to his, her or its nominees, a certificate or certificates for the number of full Class A Common Shares issuable on conversion of such Series C Preferred Shares in accordance with the provisions hereof and (2) pay cash as provided in Subsection (4.2) in lieu of any fraction of a Class A Common Share otherwise issuable upon such conversion and the payment of any declared but unpaid dividends on the Series C Preferred Shares so converted. It is intended that all Series A Preferred Shares so converted shall convert into and shall become Class B Common Shares and shall cease to exist as Series A Preferred Shares, that all Series B Preferred Shares so converted shall convert into and shall become Class A Common Shares and shall cease to exist as Series B Preferred Shares and that all Series C Preferred Shares so converted shall convert into and shall become Class A Common Shares and shall cease to exist as Series C Preferred Shares. Upon the conversion of any Preferred Share pursuant to the foregoing provisions, the number of authorised Preferred Shares, and the applicable series thereof, shall be diminished by the number of Preferred Shares that were so converted and the number of each applicable class of Common Shares shall increase in the same amount.

(5.3) Optional Conversion into Class A' Common Shares or Class B' Common Shares.

(a) Trigger Event. In connection with the IPO and notwithstanding the provisions of Subsection (5.1):

(i) Any Series A Preferred Share may be converted, in such manner as is permitted by Bermuda law, at the election of the holder thereof and in lieu of the Class B Common Share(s) otherwise issuable upon conversion of such Series A Preferred Share pursuant to Subsection (5.1), 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 Series A Conversion Price in effect at the time of such conversion. Notice of such election must be delivered to the Company at least ten (10) days prior to the anticipated closing of the IPO or such earlier time as is reasonably required by the managing underwriters(s) of the IPO. It is intended that all Series A Preferred Shares so converted shall convert into and shall become Class B' Common Shares and shall cease to exist as Series A Preferred Shares. Upon the conversion of any Series A Preferred Share pursuant to the foregoing provisions, the number of authorised Series A Preferred Shares shall be diminished by the number of Series A Preferred Shares that were so converted and the number of Class B' Common Shares shall increase in the same amount.

(ii) Any Series B Preferred Share may be converted, in such manner as is permitted by Bermuda law, at the election of the holder thereof and in lieu of the Class A Common Share(s) otherwise issuable upon conversion of such Series B Preferred Share pursuant to Subsection (5.1), 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 in effect at the time of such conversion. Notice of such election must be delivered to the Company at least ten (10) days prior to the anticipated closing of the IPO or such earlier time as is reasonably required by the managing underwriters(s) of the IPO. It is intended that all Series B Preferred Shares so converted shall convert into and shall become Class A' Common Shares and shall cease to exist as Series B Preferred Shares. Upon the conversion of any Series B Preferred Share pursuant to the foregoing provisions, the number of authorised Series B Preferred Shares shall be diminished by the number of Series B Preferred Shares that were so converted and the number of Class A' Common Shares shall increase in the same amount.

(iii) Any Series C Preferred Share may be converted, in such manner as is permitted by Bermuda law, at the election of the holder thereof and in lieu of the Class A Common Share(s) otherwise issuable upon conversion of such Series C Preferred Share pursuant to Subsection (5.1), 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 in effect at the time of such conversion. Notice of such election must be delivered to the Company at least ten (10) days prior to the anticipated closing of the IPO or such earlier time as is reasonably required by the managing underwriters(s) of the IPO. It is intended that all Series C Preferred Shares so converted shall convert into and shall become Class A' Common Shares and shall cease to exist as Series C Preferred Shares. Upon the conversion of any Series C Preferred Share pursuant to the foregoing provisions, the number of authorised Series C Preferred Shares shall be diminished by the number of Series C Preferred Shares that were so converted and the number of Class A' Common Shares shall increase in the same amount.

(b) Procedural Requirements. The provisions of Subsection (5.2) shall apply to the conversion of the Series A Preferred Shares into Class B' Common Shares pursuant to Subsection (5.3)(a)(i), the conversion of the Series B Preferred Shares into Class A' Common Shares pursuant to Subsection (5.3)(a)(ii) and the conversion of the Series C Preferred Shares into Class A' Common Shares pursuant to Subsection (5.3)(a)(iii) with such necessary changes in the details thereof as are necessitated by the context.

(c) Additional Mechanics of Conversion and Other Adjustments. The provisions of Subsections (4.2), (4.3), (4.4), (4.5), (4.6), (4.7) and (4.8) shall

apply to the conversion of the Series A Preferred Shares into Class B' Common Shares pursuant to Subsection (5.3)(a)(i), the conversion of the Series B Preferred Shares into Class A' Common Shares pursuant to Subsection (5.3)(a)(ii) and the conversion of the Series C Preferred Shares into Class A' Common Shares pursuant to Subsection (5.3)(a)(iii), with such necessary changes in the details thereof as are necessitated by the context, but without duplication of any of the rights, preferences or privileges of the applicable series of Preferred Shares otherwise provided in such subsections.

(6) Redemption. The Preferred Shares are not redeemable except in accordance with the provisions in Subsection (2.4)(b).

(7) Waiver. Unless a different vote is specified in these Bye-laws, any of the rights, powers, preferences and other terms of the Preferred Shares, Series A Preferred Shares, Series B Preferred Shares or Series C Preferred Shares set forth herein may be waived on behalf of all holders of such shares by the affirmative written consent or vote of the Requisite Holders.

4.3 Notices. Any notice required or permitted by the provisions of these Bye-laws to be given to a holder of Preferred Shares shall be mailed, postage prepaid, to the post office address last shown on the records of the Company, or given by electronic communication in compliance with the provisions of the Act, and shall be deemed sent upon such mailing or electronic transmission.

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 monies (whether in respect of nominal value or premium) unpaid on the shares allotted to or held by such Members 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 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.

5.3 The Company may accept from any Member the whole or a part of the amount remaining unpaid on any shares held by him, although no part of that amount has been called up.

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 Unless otherwise provided herein, every Member shall be entitled to a certificate under the common seal (or a facsimile thereof) of the Company 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.

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 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 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 and save as otherwise provided in these Bye-Laws) may, whether or not the Company is being wound-up, be varied with the consent in writing of the holders of at least 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 monies payable in cash in respect of the shares may be paid by cheque or 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 holder may in writing direct.

17.2 In the case of joint holders of shares, any dividend, interest or other monies payable in cash in respect of shares may be paid by cheque or 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 in writing direct. 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 monies due from such Member to the Company on account of calls or otherwise.

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 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

Subject to an election made by the Company in accordance with the Act to dispense with the holding of 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 five 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 five 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 of General Meeting

The Secretary may postpone any general meeting called in accordance with these Bye-laws (other than a meeting requisitioned under these Bye-laws) provided that notice of postponement is given to the Members before the time for such meeting. Fresh notice of the date, time and place for the postponed meeting shall be given to each Member in accordance with these Bye-laws.

25. Electronic Participation in Meetings

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.

26. Quorum at General Meetings

26.1 At any general meeting two or more persons present in person and representing in person or by proxy in excess of 50% of the voting power of the total issued voting shares in the Company throughout the meeting shall form a quorum for the transaction of business, provided that if the Company shall at any time have only one Member, one Member present in person or by proxy shall form a quorum for the transaction of business at any general meeting held during such time.

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 those attending and entitled to vote thereat, the chairman or the president of the Company, if there be one, shall act as chairman of the meeting at all general meetings at which such person is present. 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 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 An instrument appointing a proxy shall be in writing in substantially the following form or such other form as 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)

31.2 The instrument appointing 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 instrument appointing a proxy proposes to vote, and an instrument appointing a proxy which is not received in the manner so prescribed 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

The chairman of a general meeting may, with the consent of the Members at any general meeting at which a quorum is present, and shall if so directed by the meeting, adjourn the meeting. 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 The Board shall be elected or appointed at the annual general meeting or at any special general meeting called for that purpose, subject to Bye-law 4.3 D(3) 3.2, which shall also permit the appointment of certain Directors in the circumstances and manner set out therein.

36.2 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

Unless otherwise provided herein, the Board shall consist of not less than one Director or such number in excess thereof as the Members or Directors may determine.

38. Term of Office of Directors

Directors shall hold office for such term as the Members may determine or, in the absence of such determination, until the next annual general meeting or until their successors are elected or appointed or their office is otherwise vacated.

39. Alternate Directors

39.1 At any general meeting, the Members may elect a person or persons to act as a Director in the alternative to any one or more Directors or may authorise the Board to appoint such Alternate Directors.

39.2 Unless the Members otherwise resolve, any Director may appoint a person or persons to act as a Director in the alternative to himself by notice deposited with the Secretary.

39.3 Any person elected or appointed pursuant to this Bye-law shall have all the rights and powers of the Director or Directors for whom such person is elected or appointed in the alternative, provided that such person shall not be counted more than once in determining whether or not a quorum is present.

39.4 An Alternate Director shall be entitled to receive notice of all Board meetings and to attend and vote at any such meeting at which a Director for whom such Alternate Director was appointed in the alternative is not personally present and generally to perform at such meeting all the functions of such Director for whom such Alternate Director was appointed.

39.5 An Alternate Director's office shall terminate —

(a) in the case of an alternate elected by the Members:

(i) on the occurrence in relation to the Alternate Director of any event which, if it occurred in relation to the Director for whom he was elected to act, would result in the termination of that Director; or

(ii) if the Director for whom he was elected in the alternative ceases for any reason to be a Director, provided that the alternate removed in these

circumstances may be re-appointed by the Board as an alternate to the person appointed to fill the vacancy; and

(b) in the case of an alternate appointed by a Director:

(i) on the occurrence in relation to the Alternate Director of any event which, if it occurred in relation to his appointor, would result in the termination of the appointor's directorship; or

(ii) when the Alternate Director's appointor revokes the appointment by notice to the Company in writing specifying when the appointment is to terminate; or

(iii) if the Alternate Director's appointor ceases for any reason to be a Director.

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 may, at any special general meeting convened and held in accordance with these Bye-laws, remove a Director 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.

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 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 and to appoint an Alternate Director to any Director so appointed.

42. Remuneration of Directors

The remuneration (if any) of the Directors shall be determined by the Company in general meeting 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 its 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 monies 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 monies 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 monies 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. 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 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 three Directors, provided that if there is only one Director for the time being in office the quorum shall be one.

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, the chairman or the president of the Company, if there be one, shall act as chairman of the meeting at all Board meetings at which such person is present. 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. For the purposes of this Bye-law only, "the Directors" shall not include an Alternate 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, meetings of managers 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, approved or otherwise acknowledged by the Members by written resolution passed in accordance with these Bye-laws; or

(c) in circumstances where the Company has elected to dispense with the holding of an annual general meeting, be made available to the Members in accordance with the Act in such manner as the Board shall determine.

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.

VOLUNTARY WINDING-UP AND DISSOLUTION

74. 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

75. Changes to Bye-laws

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. Changes to the Memorandum of Association

No alteration or amendment to the Memorandum of Association may be made save in accordance with the Act and until same has been approved by a resolution of the Board and by a resolution of the Members.

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.

KINIKSA PHARMACEUTICALS, LTD.
SECOND AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

THIS SECOND AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT (this "**Agreement**"), is made as of February 9, 2018, by and among Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company (the "**Company**"), each of the investors listed on Schedule A hereto (the "**Investors**"), each of the Founders (as defined below), and any additional Investor that becomes a party to this Agreement in accordance with Subsection 6.9 hereof.

RECITALS

WHEREAS, the Company and certain of the Investors (the "**Prior Investors**") previously entered into an Amended and Restated Investors' Rights Agreement, dated March 8, 2017 (the "**Prior Agreement**"), in connection with the purchase of Series B Preferred Shares of the Company, par value US $0.0001 per share ("**Series B Preferred Shares**");

WHEREAS, the Prior Investors and the Company desire to induce certain other Investors to purchase Series C Preferred Shares of the Company, par value US $0.0001 per share ("**Series C Preferred Shares**"), pursuant to the Series C Preferred Shares Purchase Agreement dated as of the date hereof by and among the Company and the Investors (the "**Purchase Agreement**") by amending and restating the Prior Agreement to provide the Investors with the rights and privileges as set forth herein.

NOW, THEREFORE, the Company and the Investors, including the Prior Investors, each hereby agree to amend and restate the Prior Agreement in its entirety as set forth herein, and the parties hereto further agree as follows:

1. Definitions. For purposes of this Agreement:

1.1 "**Affiliate**" means, with respect to any specified Person, (i) any other Person who, directly or indirectly, controls, is controlled by, or is under common control with such Person, including without limitation any general partner, member, managing member, officer or director of such Person or any professional investment fund now or hereafter existing that is controlled by one or more general partners or managing members of, or shares the same management company with, such Person and (ii) such Person's Immediate Family Members or a trust for the benefit of such Person or one or more of such Person's Immediate Family Members.

1.2 "**Class A Common Shares**" means the Company's Class A common shares, par value US $0.0001 per share.

1.3 "**Class A' Common Shares**" means the Company's Class A' common shares, par value US $0.0001 per share.

1.4 "**Class B Common Shares**" means the Company's Class B common shares, par value US $0.0001 per share.

1.5 "**Class B' Common Shares**" means the Company's Class B' common shares, par value US$0.0001 per share.

1.6 "**Common Shares**" means, collectively, common shares in the Company's authorized share capital, including, without limitation, Class A Common Shares, Class A' Common Shares, Class B Common Shares and Class B' Common Shares.

1.7 "**Damages**" means any loss, damage, claim or liability (joint or several) to which a party hereto may become subject under the Securities Act, the Exchange Act, or other federal or state law, insofar as such loss, damage, claim or liability (or any action in respect thereof) arises out of or is based upon: (i) any untrue statement or alleged untrue statement of a material fact contained in any registration statement of the Company, including any preliminary prospectus or final prospectus contained therein or any amendments or supplements thereto, (ii) an omission or alleged omission to state therein a material fact required to be stated therein, or necessary to make the statements therein not misleading, or (iii) any violation or alleged violation by the indemnifying party (or any of its agents or Affiliates) of the Securities Act, the Exchange Act, any state securities law, or any rule or regulation promulgated under the Securities Act, the Exchange Act, or any state securities law.

1.8 "**Derivative Securities**" means any securities or rights convertible into, or exercisable or exchangeable for (in each case, directly or indirectly), Class A Common Shares, including options and warrants, as well as Class A' Common Shares, Class B Common Shares, Class B' Common Shares and Preferred Shares.

1.9 "**Exchange Act**" means the Securities Exchange Act of 1934, as amended, and the rules and regulations promulgated thereunder.

1.10 "**Excluded Registration**" means (i) a registration relating to the sale of securities to employees of the Company or a subsidiary pursuant to a share option, share purchase, or similar plan, (ii) a registration relating to an SEC Rule 145 transaction, (iii) a registration on any form that does not include substantially the same information as would be required to be included in a registration statement covering the sale of the Registrable Securities, or (iv) a registration in which the only Common Shares being registered are Common Shares issuable upon conversion of debt securities that are also being registered.

1.11 "**Form S-1**" means such form under the Securities Act as in effect on the date hereof or any successor registration form under the Securities Act subsequently adopted by the SEC, or any equivalent form for foreign filers, such as Form F-1.

1.12 "**Form S-3**" means such form under the Securities Act as in effect on the date hereof or any registration form under the Securities Act subsequently adopted by the SEC that permits incorporation of substantial information by reference to other documents

filed by the Company with the SEC, or any equivalent form for foreign filers, such as Form F-3.

1.13 "**Founders**" means each of those shareholders listed on Schedule B hereto; *provided* that if any such shareholder is no longer providing services to the Company or any of its subsidiaries as an officer, director, employee or consultant, such shareholder shall no longer be a Founder and Schedule B shall be updated, without requiring the consent of any party hereto, to reflect the removal of such shareholder.

1.14 "**GAAP**" means generally accepted accounting principles in the United States.

1.15 "**Holder**" means any Investor that holds Registrable Securities who is a party to this Agreement.

1.16 "**Immediate Family Member**" means a child, stepchild, grandchild, parent, stepparent, grandparent, spouse, sibling, mother-in-law, father-in-law, son-in-law, daughter-in-law, brother-in-law, or sister-in-law, including, adoptive relationships, of a natural person referred to herein.

1.17 "**Initiating Holders**" means, collectively, Holders who properly initiate a registration request under this Agreement.

1.18 "**IPO**" means the Company's first underwritten public offering of its Common Shares under the Securities Act.

1.19 "**Key Employee**" means any executive-level employee (including, division director and vice president-level positions) as well as any employee who, either alone or in concert with others, develops, invents, programs, or designs any Company Intellectual Property (as defined in the Purchase Agreement).

1.20 "**Major Investor**" means any Investor that, individually or together with such Investor's Affiliates, holds at least 1,700,000 shares of Registrable Securities on an as-converted basis (as adjusted for any share split, share dividend, combination, or other recapitalization or reclassification effected after the date hereof) (provided that any Investor that ceases to hold at least 1,700,000 such shares solely as a result of the exercise of the drag-along right of the Selling Investors (as defined in the Voting Agreement (as defined below)) on less than all of the shares held by such Investor, shall continue to be a Major Investor for the purposes of this Agreement).

1.21 "**New Securities**" means, collectively, equity securities of the Company, whether or not currently authorized, as well as rights, options, or warrants to purchase such equity securities, or securities of any type whatsoever that are, or may become, convertible or exchangeable into or exercisable for such equity securities.

1.22 "**Person**" means any individual, corporation, partnership, trust, limited liability company, association or other entity.

1.23 "**Preferred Director**" means any director of the Company that the holders of record of the Preferred Shares are entitled to elect pursuant to the Restated Bye-Laws.

1.24 "**Preferred Shares**" means, collectively, the Series A Preferred Shares of the Company, par value US $0.0001 per share, the Series B Preferred Shares and the Series C Preferred Shares.

1.25 "**Registrable Securities**" means (i) any Class A Common Shares issued or issuable (directly or indirectly) upon conversion and/or exercise of any other securities of the Company (including any Derivative Securities) held by the Investors on the date hereof or acquired by the Investors after the date hereof, and (ii) any Common Shares issued as (or issuable upon the conversion or exercise of any warrant, right, or other security that is issued as) a dividend or other distribution with respect to, or in exchange for or in replacement of, the shares referenced in clause (i) above; excluding in all cases, however, (x) any Registrable Securities sold by a Person in a transaction in which the applicable rights under this Agreement are not assigned pursuant to Subsection 6.1, and excluding, for purposes of Section 2, any shares for which registration rights have terminated pursuant to Subsection 2.13 of this Agreement and (y) any Common Shares (including only those actually issued and outstanding) held by the Investors immediately prior to the Series C Original Issue Date (as defined in the Restated Bye-Laws).

1.26 "**Registrable Securities then outstanding**" means the number of shares determined by adding the number of issued and outstanding Class A Common Shares that are Registrable Securities and the number of Class A Common Shares issuable (directly or indirectly) pursuant to then exercisable and/or convertible securities that are Registrable Securities (without respect to any period of notice that may be required to be given by the Investor to exercise or convert such securities).

1.27 "**Restricted Securities**" means the securities of the Company required to be notated with the legend set forth in Subsection 2.12(b) hereof.

1.28 "**Restated Bye-Laws**" means the Company's Amended and Restated Bye-Laws, as amended and/or restated from time to time.

1.29 "**SEC**" means the Securities and Exchange Commission.

1.30 "**SEC Rule 144**" means Rule 144 promulgated by the SEC under the Securities Act.

1.31 "**SEC Rule 145**" means Rule 145 promulgated by the SEC under the Securities Act.

1.32 "**Securities Act**" means the Securities Act of 1933, as amended, and the rules and regulations promulgated thereunder.

1.33 "**Selling Expenses**" means all underwriting discounts, selling commissions, and share transfer taxes applicable to the sale of Registrable Securities, and fees and disbursements of counsel for any Holder, except for the fees and disbursements of the Selling Holder Counsel borne and paid by the Company as provided in Subsection 2.6.

2. Registration Rights. The Company covenants and agrees as follows:

2.1 Demand Registration.

(a) Form S-1 Demand. If at any time after the earlier of (i) three years after the date of this Agreement or (ii) 180 days after the effective date of the registration statement for the IPO, the Company receives a request from Holders of at least a majority of the Registrable Securities then outstanding, on an as-converted into Class A Common Shares basis, that the Company file a Form S-1 registration statement with an anticipated aggregate offering price of not less than $10 million, then the Company shall (x) within 10 days after the date such request is given, give notice thereof (the "**Demand Notice**") to all Holders other than the Initiating Holders, and (y) as soon as practicable, and in any event within 60 days after the date such request is given by the Initiating Holders, file a Form S-1 registration statement under the Securities Act covering all Registrable Securities that the Initiating Holders requested to be registered and any additional Registrable Securities requested to be included in such registration by any other Holders, as specified by notice given by each such Holder to the Company within 10 days of the date the Demand Notice is given, and in each case, subject to the limitations of Subsections 2.1(c) and 2.3.

(b) Form S-3 Demand; Affiliate Holder Registration. If at any time when it is eligible to use a Form S-3 registration statement, the Company receives a request from Holders of at least 25% of the Registrable Securities then outstanding on an as-converted into Class A Common Shares basis that the Company file a Form S-3 registration statement with respect to issued and outstanding Registrable Securities of such Holders having an anticipated aggregate offering price of not less than $5 million, then the Company shall (i) within 10 days after the date such request is given, give a Demand Notice to all Holders other than the Initiating Holders and (ii) as soon as practicable, and in any event within 20 days after the date such request is given by the Initiating Holders, file a Form S-3 registration statement under the Securities Act covering all Registrable Securities requested to be included in such registration by any other Holders, as specified by notice given by each such Holder to the Company within 10 days of the date the Demand Notice is given, and in each case, subject to the limitations of Subsections 2.1(c) and 2.3. Notwithstanding the foregoing obligations, on the day the Company becomes eligible to use a Form S-3 registration statement (or the next business days thereafter if such day is not a business day), the Company shall, without obligation of any Demand Notice being provided by a Holder, file a Form S-3 registration statement under the Securities Act covering all Registrable Securities held by any Holders who at the time would be considered an "affiliate" of the Company under SEC Rule 144 ("**Affiliate Holders**").

(c) Notwithstanding the foregoing obligations, if the Company furnishes to Holders requesting a registration pursuant to this Subsection 2.1 and/or the Affiliate Holders who are otherwise entitled to a registration under Subsection 2.1(b) a certificate signed

by the Company's chief executive officer stating that, in the good faith judgment of the Company's Board of Directors, it would be materially detrimental to the Company and its shareholders for such registration statement to either become effective or remain effective for as long as such registration statement otherwise would be required to remain effective because such action would (i) materially interfere with a significant acquisition, corporate reorganization, or other similar transaction involving the Company, (ii) require premature disclosure of material information that the Company has a bona fide business purpose for preserving as confidential, or (iii) render the Company unable to comply with requirements under the Securities Act or Exchange Act, then the Company shall have the right to defer taking action with respect to such filing, and any time periods with respect to filing or effectiveness thereof shall be tolled correspondingly, for a period of not more than 90 days after the request of the Initiating Holders is given; provided, however, that the Company may not invoke this right more than once in any 12-month period; and provided further that the Company shall not register any securities for its own account or that of any other shareholder during such 90-day period other than an Excluded Registration.

(d) The Company shall not be obligated to effect, or to take any action to effect, any registration pursuant to Subsection 2.1(a)(i) during the period that is 45 days before the Company's good faith estimate of the date of filing of, and ending on a date that is 180 days after the effective date of, a Company-initiated registration, provided that the Company is actively employing in good faith commercially reasonable efforts to cause such registration statement to become effective, (ii) after the Company has effected two registrations pursuant to Subsection 2.1(a), or (iii) if the Initiating Holders propose to dispose of Registrable Securities that may be immediately registered on Form S-3 pursuant to a request made pursuant to Subsection 2.1(b). The Company shall not be obligated to effect, or to take any action to effect, any registration pursuant to Subsection 2.1(b): (i) during the period that is 30 days before the Company's good faith estimate of the date of filing of, and ending on a date that is 90 days after the effective date of, a Company-initiated registration, provided that the Company is actively employing in good faith commercially reasonable efforts to cause such registration statement to become effective, or (ii) if the Company has effected two registrations pursuant to Subsection 2.1(b) within the 12-month period immediately preceding the date of such request. A registration shall not be counted as "effected" for purposes of this Subsection 2.1(d) until such time as the applicable registration statement has been declared effective by the SEC and all Registrable Securities requested to be registered have been so registered, unless the Initiating Holders withdraw their request for such registration, elect not to pay the registration expenses therefor and forfeit their right to one demand registration statement pursuant to Subsection 2.6; provided, however, that if the request to withdraw such registration is made as a result of a material adverse change of the Company, such withdrawn registration statement shall not be counted as "effected" for purposes of this Subsection 2.1(d).

2.2 Company Registration. If the Company proposes to register (including, for this purpose, a registration effected by the Company for shareholders other than the Holders) any of its securities under the Securities Act in connection with the public offering of such securities solely for cash (other than in an Excluded Registration), the Company shall, at such time, promptly give each Holder notice of such registration. Upon the request of each Holder given within 10 days after such notice is given by the Company, the Company shall, subject to

the provisions of Subsection 2.3, cause to be registered all of the Registrable Securities that each such Holder has requested to be included in such registration. The Company shall have the right to terminate or withdraw any registration initiated by it under this Subsection 2.2 before the effective date of

such registration, whether or not any Holder has elected to include Registrable Securities in such registration. The expenses (other than Selling Expenses) of such withdrawn registration shall be borne by the Company in accordance with Subsection 2.6.

2.3 Underwriting Requirements.

(a) If, pursuant to Subsection 2.1, the Initiating Holders intend to distribute the Registrable Securities covered by their request by means of an underwriting, they shall so advise the Company as a part of their request made pursuant to Subsection 2.1, and the Company shall include such information in the Demand Notice. The underwriter(s) will be selected by the Initiating Holders, subject only to the reasonable approval of the Company. In such event, the right of any Holder to include such Holder's Registrable Securities in such registration shall be conditioned upon such Holder's participation in such underwriting and the inclusion of such Holder's Registrable Securities in the underwriting to the extent provided herein. All Holders proposing to distribute their securities through such underwriting shall, together with the Company as provided in Subsection 2.4(e), enter into an underwriting agreement in customary form with the underwriter(s) selected for such underwriting. Notwithstanding any other provision of this Subsection 2.3, if the managing underwriter(s) advise(s) the Initiating Holders in writing that marketing factors require a limitation on the number of shares to be underwritten, then the Initiating Holders shall so advise all Holders of Registrable Securities that otherwise would be underwritten pursuant hereto, and the number of Registrable Securities that may be included in the underwriting shall be allocated among such Holders of Registrable Securities, including the Initiating Holders, in proportion (as nearly as practicable) to the number of Registrable Securities owned by each Holder or in such other proportion as shall mutually be agreed to by all such selling Holders; provided, however, that the number of Registrable Securities held by the Holders to be included in such underwriting shall not be reduced unless all other securities are first entirely excluded from the underwriting. To facilitate the allocation of shares in accordance with the above provisions, the Company or the underwriters may round the number of shares allocated to any Holder to the nearest one hundred (100) shares.

(b) In connection with any offering involving an underwriting of the Company's share capital pursuant to Subsection 2.2, the Company shall not be required to include any of the Holders' Registrable Securities in such underwriting unless the Holders accept the terms of the underwriting as agreed upon between the Company and its underwriters, and then only in such quantity as the underwriters in their sole discretion determine will not jeopardize the success of the offering by the Company. If the total number of securities, including Registrable Securities, requested by shareholders to be included in such offering exceeds the number of securities to be sold (other than by the Company) that the underwriters in their reasonable discretion determine is compatible with the success of the offering, then the Company shall be required to include in the offering only that number of such securities, including Registrable Securities, which the underwriters and the Company in their sole discretion determine will not jeopardize the success of the offering. If the underwriters determine that less

than all of the Registrable Securities requested to be registered can be included in such offering, then the Registrable Securities that are included in such offering shall be allocated among the selling Holders in proportion (as nearly as practicable to) the number of Registrable Securities owned by each selling Holder or in such other proportions as shall mutually be agreed to by all such selling Holders. To facilitate the allocation of shares in accordance with the above provisions, the Company or the underwriters may round the number of shares allocated to any Holder to the nearest 100 shares. Notwithstanding the foregoing, in no event shall (i) the number of Registrable Securities included in the offering be reduced unless all other securities (other than securities to be sold by the Company) are first entirely excluded from the offering, or (ii) the number of Registrable Securities included in the offering be reduced below 25% of the total number of securities included in such offering, unless such offering is the IPO, in which case the selling Holders may be excluded further if the underwriters make the determination described above and no other shareholder's securities are included in such offering. For purposes of the provision in this Subsection 2.3(b) concerning apportionment, for any selling Holder that is a partnership, limited liability company, or corporation, the partners, members, retired partners, retired members, shareholders, and Affiliates of such Holder, or the estates and Immediate Family Members of any such partners, retired partners, members, and retired members and any trusts for the benefit of any of the foregoing Persons, shall be deemed to be a single "selling Holder," and any pro rata reduction with respect to such "selling Holder" shall be based upon the aggregate number of Registrable Securities owned by all Persons included in such "selling Holder," as defined in this sentence.

2.4 Obligations of the Company. Whenever required under this Section 2 to effect the registration of any Registrable Securities, the Company shall, as expeditiously as reasonably possible:

(a) prepare and file with the SEC a registration statement with respect to such Registrable Securities and use its commercially reasonable efforts to cause such registration statement to become effective and, upon the request of the Holders of a majority of the Registrable Securities registered thereunder, keep such registration statement effective for a period of up to 120 days or, if earlier, until the distribution contemplated in the registration statement has been completed; provided, however, that (i) such 120-day period shall be extended for a period of time equal to the period the Holder refrains, at the request of an underwriter of Common Shares (or other securities) of the Company, from selling any securities included in such registration, and (ii) in the case of any registration of Registrable Securities on Form S-3 that are intended to be offered on a continuous or delayed basis, subject to compliance with applicable SEC rules, such 120-day period shall be extended, if necessary, to keep the registration statement effective until all such Registrable Securities are sold;

(b) prepare and file with the SEC such amendments and supplements to such registration statement, and the prospectus used in connection with such registration statement, as may be necessary to comply with the Securities Act in order to enable the disposition of all securities covered by such registration statement;

(c) furnish to the selling Holders such numbers of copies of a prospectus, including a preliminary prospectus, as required by the Securities Act, and such other

documents as the Holders may reasonably request in order to facilitate their disposition of their Registrable Securities;

(d) use its commercially reasonable efforts to register and qualify the securities covered by such registration statement under such other securities or blue-sky laws of such jurisdictions as shall be reasonably requested by the selling Holders; provided that the Company shall not be

required to qualify to do business or to file a general consent to service of process in any such states or jurisdictions, unless the Company is already subject to service in such jurisdiction and except as may be required by the Securities Act;

(e) in the event of any underwritten public offering, enter into and perform its obligations under an underwriting agreement, in usual and customary form, with the underwriter(s) of such offering;

(f) use its commercially reasonable efforts to cause all such Registrable Securities covered by such registration statement to be listed on a national securities exchange or trading system and each securities exchange and trading system (if any) on which similar securities issued by the Company are then listed;

(g) provide a transfer agent and registrar for all Registrable Securities registered pursuant to this Agreement and provide a CUSIP number for all such Registrable Securities, in each case not later than the effective date of such registration;

(h) promptly make available for inspection by the selling Holders, any managing underwriter(s) participating in any disposition pursuant to such registration statement, and any attorney or accountant or other agent retained by any such underwriter or selected by the selling Holders, all financial and other records, pertinent corporate documents, and properties of the Company, and cause the Company's officers, directors, employees, and independent accountants to supply all information reasonably requested by any such seller, underwriter, attorney, accountant, or agent, in each case, as necessary or advisable to verify the accuracy of the information in such registration statement and to conduct appropriate due diligence in connection therewith;

(i) notify each selling Holder, promptly after the Company receives notice thereof, of the time when such registration statement has been declared effective or a supplement to any prospectus forming a part of such registration statement has been filed; and

(j) after such registration statement becomes effective, notify each selling Holder of any request by the SEC that the Company amend or supplement such registration statement or prospectus.

In addition, the Company shall ensure that, at all times after any registration statement covering a public offering of securities of the Company under the Securities Act shall have become effective, its insider trading policy shall provide that the Company's directors may implement a trading program under Rule 10b5-1 of the Exchange Act.

2.5 Furnish Information. It shall be a condition precedent to the obligations of the Company to take any action pursuant to this Section 2 with respect to the Registrable Securities of any selling Holder that such Holder shall furnish to the Company such information regarding itself, the Registrable Securities held by it, and the intended method of disposition of such securities as is reasonably required to effect the registration of such Holder's Registrable Securities.

2.6 Expenses of Registration. All expenses (other than Selling Expenses) incurred in connection with registrations, filings, or qualifications pursuant to Section 2, including all registration, filing, and qualification fees; printers' and accounting fees; fees and disbursements of counsel for the Company; and the reasonable fees and disbursements, not to exceed $50,000, of one counsel for the selling Holders ("**Selling Holder Counsel**"), shall be borne and paid by the Company; provided, however, that the Company shall not be required to pay for any expenses of any registration proceeding begun pursuant to Subsection 2.1 if the registration request is subsequently withdrawn at the request of the Holders of a majority of the Registrable Securities to be registered (in which case all selling Holders shall bear such expenses pro rata based upon the number of Registrable Securities that were to be included in the withdrawn registration), unless the Holders of a majority of the Registrable Securities agree to forfeit their right to one registration pursuant to Subsections 2.1(a) or 2.1(b), as the case may be; provided further that if, at the time of such withdrawal, the Holders shall have learned of a material adverse change in the condition, business, or prospects of the Company from that known to the Holders at the time of their request and have withdrawn the request with reasonable promptness after learning of such information then the Holders shall not be required to pay any of such expenses and shall not forfeit their right to one registration pursuant to Subsections 2.1(a) or 2.1(b). All Selling Expenses relating to Registrable Securities registered pursuant to this Section 2 shall be borne and paid by the Holders pro rata on the basis of the number of Registrable Securities registered on their behalf.

2.7 Delay of Registration. No Holder shall have any right to obtain or seek an injunction restraining or otherwise delaying any registration pursuant to this Agreement as the result of any controversy that might arise with respect to the interpretation or implementation of this Section 2.

2.8 Indemnification. If any Registrable Securities are included in a registration statement under this Section 2:

(a) To the extent permitted by law, the Company will indemnify and hold harmless each selling Holder, and the partners, members, officers, directors, and shareholders of each such Holder; legal counsel and accountants for each such Holder; any underwriter (as defined in the Securities Act) for each such Holder; and each Person, if any, who controls such Holder or underwriter within the meaning of the Securities Act or the Exchange Act, against any Damages, and the Company will pay to each such Holder, underwriter, controlling Person, or other aforementioned Person any legal or other expenses reasonably incurred thereby in connection with investigating or defending any claim or proceeding from which Damages may result, as such expenses are incurred; provided, however, that the indemnity agreement contained in this Subsection 2.8(a) shall not apply to amounts paid in settlement of

any such claim or proceeding if such settlement is effected without the consent of the Company, which consent shall not be unreasonably withheld, nor shall the Company be liable for any Damages to the extent that they arise out of or are based upon actions or omissions made in reliance upon and in conformity with written information furnished by or on behalf of any such Holder, underwriter, controlling Person, or other aforementioned Person expressly for use in connection with such registration.

(b) To the extent permitted by law, each selling Holder, severally and not jointly, will indemnify and hold harmless the Company, and each of its directors, each of its officers who has signed the registration statement, each Person (if any), who controls the Company within the meaning of the Securities Act, legal counsel and accountants for the Company, any underwriter (as defined in the Securities Act), any other Holder selling securities in such registration statement, and any controlling Person of any such underwriter or other Holder, against any Damages, in each case only to the extent that such Damages arise out of or are based upon actions or omissions made in reliance upon and in conformity with written information furnished by or on behalf of such selling Holder expressly for use in connection with such registration; and each such selling Holder will pay to the Company and each other aforementioned Person any legal or other expenses reasonably incurred thereby in connection with investigating or defending any claim or proceeding from which Damages may result, as such expenses are incurred; provided, however, that the indemnity agreement contained in this Subsection 2.8(b) shall not apply to amounts paid in settlement of any such claim or proceeding if such settlement is effected without the consent of the Holder, which consent shall not be unreasonably withheld; and provided further that in no event shall the aggregate amounts payable by any Holder by way of indemnity or contribution under Subsections 2.8(b) and 2.8(d) exceed the proceeds from the offering received by such Holder (net of any Selling Expenses paid by such Holder), except in the case of fraud or willful misconduct by such Holder.

(c) Promptly after receipt by an indemnified party under this Subsection 2.8 of notice of the commencement of any action (including any governmental action) for which a party may be entitled to indemnification hereunder, such indemnified party will, if a claim in respect thereof is to be made against any indemnifying party under this Subsection 2.8, give the indemnifying party notice of the commencement thereof. The indemnifying party shall have the right to participate in such action and, to the extent the indemnifying party so desires, participate jointly with any other indemnifying party to which notice has been given, and to assume the defense thereof with counsel mutually satisfactory to the parties; provided, however, that an indemnified party (together with all other indemnified parties that may be represented without conflict by one counsel) shall have the right to retain one separate counsel, with the fees and expenses to be paid by the indemnifying party, if representation of such indemnified party by the counsel retained by the indemnifying party would be inappropriate due to actual or potential differing interests between such indemnified party and any other party represented by such counsel in such action. The failure to give notice to the indemnifying party within a reasonable time of the commencement of any such action shall relieve such indemnifying party of any liability to the indemnified party under this Subsection 2.8, to the extent that such failure materially prejudices the indemnifying party's ability to defend such action. The failure to give notice to the indemnifying party will not relieve

it of any liability that it may have to any indemnified party otherwise than under this Subsection 2.8.

(d) To provide for just and equitable contribution to joint liability under the Securities Act in any case in which either: (i) any party otherwise entitled to indemnification hereunder makes a claim for indemnification pursuant to this Subsection 2.8 but it is judicially determined (by the entry of a final judgment or decree by a court of competent jurisdiction and the expiration of time to appeal or the denial of the last right of appeal) that such indemnification may not be enforced in such case, notwithstanding the fact that this Subsection 2.8 provides for indemnification in such case, or (ii) contribution under the Securities Act may be required on the part of any party hereto for which indemnification is provided under this Subsection 2.8, then, and in each such case, such parties will contribute to the aggregate losses, claims, damages, liabilities, or expenses to which they may be subject (after contribution from others) in such proportion as is appropriate to reflect the relative fault of each of the indemnifying party and the indemnified party in connection with the statements, omissions, or other actions that resulted in such loss, claim, damage, liability, or expense, as well as to reflect any other relevant equitable considerations. The relative fault of the indemnifying party and of the indemnified party shall be determined by reference to, among other things, whether the untrue or allegedly untrue statement of a material fact, or the omission or alleged omission of a material fact, relates to information supplied by the indemnifying party or by the indemnified party and the parties' relative intent, knowledge, access to information, and opportunity to correct or prevent such statement or omission; provided, however, that, in any such case (x) no Holder will be required to contribute any amount in excess of the public offering price of all such Registrable Securities offered and sold by such Holder pursuant to such registration statement, and (y) no Person guilty of fraudulent misrepresentation (within the meaning of Section 11(f) of the Securities Act) will be entitled to contribution from any Person who was not guilty of such fraudulent misrepresentation; and provided further that in no event shall a Holder's liability pursuant to this Subsection 2.8(d), when combined with the amounts paid or payable by such Holder pursuant to Subsection 2.8(b), exceed the proceeds from the offering received by such Holder (net of any Selling Expenses paid by such Holder), except in the case of willful misconduct or fraud by such Holder.

(e) Notwithstanding the foregoing, to the extent that the provisions on indemnification and contribution contained in the underwriting agreement entered into in connection with the underwritten public offering are in conflict with the foregoing provisions, the provisions in the underwriting agreement shall control.

(f) Unless otherwise superseded by an underwriting agreement entered into in connection with the underwritten public offering, the obligations of the Company and Holders under this Subsection 2.8 shall survive the completion of any offering of Registrable Securities in a registration under this Section 2, and otherwise shall survive the termination of this Agreement.

2.9 Reports Under Exchange Act. With a view to making available to the Holders the benefits of SEC Rule 144 and any other rule or regulation of the SEC that may at

any time permit a Holder to sell securities of the Company to the public without registration or pursuant to a registration on Form S-3, the Company shall:

(a) make and keep available adequate current public information, as those terms are understood and defined in SEC Rule 144, at all times after the effective date of the registration statement filed by the Company for the IPO;

(b) use commercially reasonable efforts to file with the SEC in a timely manner all reports and other documents required of the Company under the Securities Act and the Exchange Act (at any time after the Company has become subject to such reporting requirements); and

(c) furnish to any Holder, so long as the Holder owns any Registrable Securities, forthwith upon request (i) to the extent accurate, a written statement by the Company that it has complied with the reporting requirements of SEC Rule 144 (at any time after 90 days after the effective date of the registration statement filed by the Company for the IPO), the Securities Act, and the Exchange Act (at any time after the Company has become subject to such reporting requirements), or that it qualifies as a registrant whose securities may be resold pursuant to Form S-3 (at any time after the

Company so qualifies), and (ii) such other information as may be reasonably requested in availing any Holder of any rule or regulation of the SEC that permits the selling of any such securities without registration (at any time after the Company has become subject to the reporting requirements under the Exchange Act) or pursuant to Form S-3 (at any time after the Company so qualifies to use such form).

2.10 Limitations on Subsequent Registration Rights. From and after the date of this Agreement, the Company shall not, without the prior written consent of the Holders of a majority of the Registrable Securities then outstanding, enter into any agreement with any holder or prospective holder of any securities of the Company that (i) would allow such holder or prospective holder to include such securities in any registration unless, under the terms of such agreement, such holder or prospective holder may include such securities in any such registration only to the extent that the inclusion of such securities will not reduce the number of the Registrable Securities of the Holders that are included, or (ii) would allow such holder or prospective holder to initiate a demand for registration of any securities held by such holder or prospective holder; provided that this limitation shall not apply to any additional Investor who becomes a party to this Agreement in accordance with Subsection 6.9 with respect to the registration rights afforded hereunder.

2.11 Lock-Up Agreement. If requested by the managing underwriter in connection with the IPO, each Holder hereby agrees that it will not, without the prior written consent of the managing underwriter, during the period commencing on the date of the final prospectus relating to the registration by the Company of any portion of its share capital and ending on the date specified by the Company and the managing underwriter (such period not to exceed 180 days): (i) lend; offer; pledge; sell; contract to sell; sell any option or contract to purchase; purchase any option or contract to sell; grant any option, right, or warrant to purchase; or otherwise transfer or dispose of, directly or indirectly, any Class A Common Shares or any securities convertible into or exercisable or exchangeable (directly or indirectly) for Class A

Common Shares held immediately before the effective date of the registration statement for such offering, or (ii) enter into any swap or other arrangement that transfers to another, in whole or in part, any of the economic consequences of ownership of such securities, whether any such transaction described in clause (i) or (ii) above is to be settled by delivery of Common Shares or other securities, in cash, or otherwise. The foregoing provisions of this Subsection 2.11 shall not apply to: (x) any Class A Common Shares purchased by a Holder in the subject offering or at any time subsequent to the subject offering, (y) the sale of any shares to an underwriter pursuant to an underwriting agreement, or (z) the transfer of any shares to any trust for the direct or indirect benefit of the Holder or the immediate family of the Holder, provided that the trustee of the trust agrees to be bound in writing by the restrictions set forth herein, and provided further that any such transfer shall not involve a disposition for value, and shall be applicable to the Holders only if all officers and directors are subject to the same restrictions and the Company obtains a similar agreement from all shareholders individually owning more than 1% of the Company's issued and outstanding Common Shares (after giving effect to conversion into Common Shares of all issued and outstanding Preferred Shares). The underwriters in connection with such registration are intended third-party beneficiaries of this Subsection 2.11 and shall have the right, power and authority to enforce the provisions hereof as though they were a party hereto. Each Holder further agrees to execute such agreements as may be reasonably requested by the underwriters in connection with such registration that are consistent with this Subsection 2.11 or that are necessary to give further effect thereto. Any discretionary waiver or termination of the restrictions of any or all of such agreements by the Company or the underwriters shall apply pro rata to all Holders subject to such agreements, based on the number of shares subject to such agreements.

2.12 Restrictions on Transfer.

(a) The Preferred Shares, the Common Shares and the Registrable Securities shall not be sold, pledged, or otherwise transferred, and the Company shall not recognize and shall issue stop-transfer instructions to its transfer agent with respect to any such sale, pledge, or transfer, except upon the conditions specified in this Agreement, which conditions are intended to ensure compliance with the provisions of the Securities Act. A transferring Holder will cause any proposed purchaser, pledgee, or transferee of Preferred Shares, Common Shares and/or Registrable Securities held by such Holder, as applicable, to agree to take and hold such securities subject to the provisions and upon the conditions specified in this Agreement.

(b) Each certificate, instrument, or book entry representing an Investor's (i) Preferred Shares, (ii) Common Shares, (iii) Registrable Securities and (iv) any other securities issued in respect of the securities referenced in clauses (i), (ii) and (iii), upon any share split, share dividend, recapitalization, merger, consolidation, or similar event, shall (unless otherwise permitted by the provisions of Subsection 2.12(c)) be notated with a legend substantially in the following form:

THE SECURITIES REPRESENTED HEREBY HAVE BEEN ACQUIRED FOR INVESTMENT AND HAVE NOT BEEN REGISTERED UNDER THE SECURITIES ACT OF 1933. SUCH SHARES MAY NOT BE SOLD,

PLEDGED, OR TRANSFERRED IN THE ABSENCE OF SUCH REGISTRATION OR A VALID EXEMPTION FROM THE REGISTRATION AND PROSPECTUS DELIVERY REQUIREMENTS OF SAID ACT.

THE SECURITIES REPRESENTED HEREBY MAY BE TRANSFERRED ONLY IN ACCORDANCE WITH THE TERMS OF AN AGREEMENT BETWEEN THE COMPANY AND THE SHAREHOLDER, A COPY OF WHICH IS ON FILE WITH THE SECRETARY OF THE COMPANY.

The holders of such Restricted Securities consent to the Company making a notation in its records and giving instructions to any transfer agent of the Restricted Securities in order to implement the restrictions on transfer set forth in this Subsection 2.12.

(c) The holder of such Restricted Securities, by acceptance of ownership thereof, agrees to comply in all respects with the provisions of this Section 2. Before any proposed sale, pledge, or transfer of any Restricted Securities, unless there is in effect a registration statement under the Securities Act covering the proposed transaction, the holder thereof shall give notice to the Company of such holder's intention to effect such sale, pledge, or transfer. Each such notice shall describe the manner and circumstances of the proposed sale, pledge, or transfer in sufficient detail and, if reasonably requested by the Company, shall be accompanied at such holder's expense by either (i) a written opinion of legal counsel who shall, and whose legal opinion shall, be reasonably satisfactory to the Company, addressed to the Company, to the effect that the proposed transaction may be effected without registration under the Securities Act, (ii) a "no action" letter from the SEC to the effect that the proposed sale, pledge, or transfer of such Restricted Securities without registration will not result in a recommendation by the staff of the SEC that action be taken with respect thereto, or (iii) any other evidence reasonably

satisfactory to counsel to the Company to the effect that the proposed sale, pledge, or transfer of the Restricted Securities may be effected without registration under the Securities Act, whereupon the holder of such Restricted Securities shall be entitled to sell, pledge, or transfer such Restricted Securities in accordance with the terms of the notice given by the holder to the Company. The Company will not require such a legal opinion or "no action" letter (x) in any transaction in compliance with SEC Rule 144, or (y) in any transaction in which such holder distributes Restricted Securities to an Affiliate of such holder for no consideration; provided that each transferee agrees in writing to be subject to the terms of this Subsection 2.12. Each certificate, instrument, or book entry representing the Restricted Securities transferred as above provided shall be notated with, except if such transfer is made pursuant to SEC Rule 144, the appropriate restrictive legend set forth in Subsection 2.12(b), except that such certificate instrument, or book entry shall not be notated with such restrictive legend if, in the opinion of counsel for such holder and the Company, such legend is not required in order to establish compliance with any provisions of the Securities Act.

2.13 Termination of Registration Rights. The right of any Holder to request registration or inclusion of Registrable Securities in any registration pursuant to Subsections 2.1 or 2.2 shall terminate upon (a) the closing of a Deemed Liquidation Event, as such term is defined in the Restated Bye-Laws; or (b) such time as SEC Rule 144 would permit sale of all of such Holder's shares without restriction.

3. Information Rights.

3.1 Delivery of Financial Statements. The Company shall deliver to each Major Investor:

(a) as soon as practicable, but in any event within 180 days after the end of each fiscal year of the Company (i) a balance sheet as of the end of such year, (ii) statements of income and of cash flows for such year, and (iii) a statement of shareholders' equity as of the end of such year, all such financial statements audited and certified by independent public accountants of recognized standing selected by the Company;

(b) as soon as practicable, but in any event within 40 days after the end of each quarter of each fiscal year of the Company, unaudited statements of income and cash flows for such fiscal quarter, and an unaudited balance sheet and a statement of shareholders' equity as of the end of such fiscal quarter, all prepared in accordance with GAAP (except that such financial statements may (i) be subject to normal year-end audit adjustments, and (ii) not contain all notes thereto that may be required in accordance with GAAP);

(c) as soon as practicable, but in any event within 40 days after the end of each of the first three quarters of each fiscal year of the Company, a statement showing the number of shares of each class and series of the Company's share capital and securities convertible into or exercisable for shares in the Company's share capital outstanding at the end of the period, the Common Shares issuable upon conversion or exercise of any issued and outstanding securities convertible or exercisable for Common Shares and the exchange ratio or exercise price applicable thereto, and the number of issued share options and share options not yet issued but reserved for issuance, if any, all in sufficient detail as to permit the Major Investors to calculate their respective percentage equity ownership in the Company, and certified by the chief financial officer or chief executive officer of the Company as being true, complete and correct;

(d) as soon as practicable, but in any event within 45 days of the end of each year, the tax information required of the Investors under United States tax law related to the Company's foreign residence, including, but not limited to, the Company's status for United States federal income tax purposes as a "passive foreign investment company."

(e) as soon as practicable, but in any event 7 days before the end of each fiscal year, a budget for the next fiscal year (collectively, the "**Budget**"), presented to the Board of Directors, including balance sheets, income statements, and statements of cash flow and, promptly after prepared, any other budgets or revised budgets prepared by the Company; and

(f) such other information relating to the financial condition, business, prospects, or corporate affairs of the Company as any Major Investor may from time to time reasonably request; provided, however, that the Company shall not be obligated under this Subsection 3.1 to provide information (i) that the Company reasonably determines in good faith to be a trade secret or confidential information (unless covered by an enforceable confidentiality

agreement, in a form acceptable to the Company) or (ii) the disclosure of which would adversely affect the attorney-client privilege between the Company and its counsel.

If, for any period, the Company has any subsidiary whose accounts are consolidated with those of the Company, then in respect of such period the financial statements delivered pursuant to the foregoing sections shall be the consolidated and consolidating financial statements of the Company and all such consolidated subsidiaries.

Notwithstanding anything else in this Subsection 3.1 to the contrary, the Company may cease providing the information set forth in this Subsection 3.1 during the period starting with the date 30 days before the Company's good-faith estimate of the date of filing of a registration statement if it reasonably concludes it must do so to comply with the SEC rules applicable to such registration statement and related offering; provided that the Company's covenants under this Subsection 3.1 shall be reinstated at such time as the Company is no longer actively employing its commercially reasonable efforts to cause such registration statement to become effective.

3.2 Inspection. The Company shall permit each Major Investor, at such Major Investor's expense, to visit and inspect the Company's properties; examine its books of account and records; and discuss the Company's affairs, finances, and accounts with its officers, during normal business hours of the Company as may be reasonably requested by the Major Investor; provided, however, that the Company shall not be obligated pursuant to this Subsection 3.2 to provide access to any information that it reasonably and in good faith considers to be a trade secret or confidential information (unless covered by an enforceable confidentiality agreement, in form acceptable to the Company) or the disclosure of which would adversely affect the attorney-client privilege between the Company and its counsel.

3.3 Termination of Information. The covenants set forth in Subsection 3.1 and Subsection 3.2 shall terminate and be of no further force or effect (i) immediately before the consummation of the IPO, (ii) when the Company becomes subject to the periodic reporting requirements of Section 12(g) or 15(d) of the Exchange Act, or (iii) upon a Deemed Liquidation Event, as such term is defined in the Restated Bye-laws, whichever event occurs first.

3.4 Confidentiality. Each Investor agrees that such Investor will keep confidential and will not disclose or divulge any confidential information obtained from the Company pursuant to the terms of this Agreement (including notice of the Company's intention to file a registration statement), unless such confidential information (a) is known or becomes known to the public in general (other than as a result of a breach of this Subsection 3.4 by such Investor), (b) is or has been independently developed or conceived by the Investor without use of the Company's confidential information, or (c) is or has been made known or disclosed to the Investor by a third party without a breach of any obligation of confidentiality such third party may have to the Company; provided, however, that an Investor may disclose confidential information (i) to its attorneys, accountants, consultants, and other professionals to the extent necessary to obtain their services in connection with monitoring its investment in the Company, (ii) to any prospective purchaser of any Registrable Securities from such Investor, if such prospective purchaser agrees to be bound by the provisions of this Subsection 3.4, (iii) to any

Affiliate, partner, limited partner, member, shareholder, or wholly owned subsidiary of such Investor or its Affiliates in the ordinary course of business, provided that such Investor informs such Person that such information is confidential and directs such Person to maintain the confidentiality of such information, or (iv) as may otherwise be required by law, provided that the Investor promptly notifies the Company of such disclosure and takes reasonable steps to minimize the extent of any such required disclosure.

4. Rights to Future Share Issuances.

4.1 Right of First Offer. Subject to the terms and conditions of this Subsection 4.1 and applicable securities laws, if the Company proposes to offer or sell any New Securities, the Company shall first offer such New Securities to each Holder and each Founder. Each Holder and each Founder shall be entitled to apportion the right of first offer hereby granted to it, in such proportions as it deems appropriate, among (i) itself, (ii) its Affiliates and (iii) its beneficial interest holders, such as limited partners, members, or any other Person having "beneficial ownership," as such term is defined in Rule 13d-3 promulgated under the Exchange Act, of such Holder ("**Investor Beneficial Owners**"); provided that each such Affiliate or Investor Beneficial Owner agrees to enter into this Agreement and each of the Voting Agreement and Second Amended and Restated Right of First Refusal and Co-Sale Agreement of even date herewith among the Company, the Investors and the other parties named therein, as an "Investor" under each such agreement.

(a) The Company shall give notice (the "**Offer Notice**") to each Holder and Founder, stating (i) its bona fide intention to offer such New Securities, (ii) the number of such New Securities to be offered, and (iii) the price and terms, if any, upon which it proposes to offer such New Securities.

(b) By notification to the Company within 10 days after the Offer Notice is given, each Holder and Founder may elect to purchase or otherwise acquire, at the price and on the terms specified in the Offer Notice, up to that portion of such New Securities (without duplication for any Holder that is also a Founder) which equals the proportion that the Class A Common Shares then held by such Holder or Founder (including all Class A Common Shares then issuable (directly or indirectly) upon conversion and/or exercise, as applicable, of the Preferred Shares and any other Derivative Securities then held by such Holder or Founder) bears to the total Class A Common Shares of the Company then issued and outstanding (assuming full conversion and/or exercise, as applicable, of all Preferred Shares and other Derivative Securities (directly or indirectly) into Class A Common Shares). At the expiration of such 10-day period, the Company shall promptly notify each Holder that has elected to purchase or acquire all the shares available to it (each, a "**Fully Exercising Holder**") and each Founder that elects to purchase or acquire all the shares available to it (each, a "**Fully Exercising Founder**") of any other Holder's or Founder's, as the case may be, failure to do likewise. During the 10-day period commencing after the Company has given such notice, each Fully Exercising Holder and Fully Exercising Founder may, by giving notice to the Company, elect to purchase or acquire, in addition to the number of shares specified above, up to that portion of the New Securities for which Holders or Founders were entitled to subscribe but that were not subscribed for by the Holders or Founders which is equal to the proportion that the Class A Common Shares issued

and held, or issuable (directly or indirectly) upon conversion and/or exercise, as applicable, of Preferred Shares and any other Derivative Securities then held, by such Fully Exercising Holder or Fully Exercising Founder bears to the Class A Common Shares issued and held, or issuable (directly or indirectly) upon conversion and/or exercise, as applicable, of the Preferred Shares and any other Derivative Securities then held, by all Fully Exercising Holders and/or Fully Exercising Founders who wish to purchase such unsubscribed shares. The closing of any sale pursuant to this Subsection 4.1(b) shall occur within the later of 90 days of the date that the Offer Notice is given and the date of initial sale of New Securities pursuant to Subsection 4.1(c).

(c) If all New Securities referred to in the Offer Notice are not elected to be purchased or acquired as provided in Subsection 4.1(b), the Company may, during the 90-day period following the expiration of the periods provided in Subsection 4.1(b), offer and sell the remaining unsubscribed portion of such New Securities to any Person or Persons at a price not less than, and upon terms no more favorable to the offeree than, those specified in the Offer Notice.

(d) The right of first offer in this Subsection 4.1 shall not be applicable to: (i) Exempted Securities (as defined in the Restated Bye-Laws); (ii) any Common Shares issued in the IPO; and (iii) any Preferred Shares issued pursuant to the Purchase Agreement.

4.2 Termination. The covenants set forth in Subsection 4.1 and Subsection 4.2 shall terminate and be of no further force or effect (i) immediately before the consummation of the IPO, or (ii) upon a Deemed Liquidation Event, as such term is defined in the Restated Bye-Laws, whichever event occurs first.

5. Additional Covenants.

5.1 Insurance. The Company shall use commercially reasonable efforts to maintain from financially sound and reputable insurers Directors and Officers liability insurance in an amount and on terms and conditions satisfactory to the Board of Directors until such time as the Board of

Directors determines that such insurance should be discontinued.

5.2 Employee Agreements and Share Vesting. The Company will cause (a) each person now or hereafter employed by it or by any subsidiary (or engaged by the Company or any subsidiary as a consultant/independent contractor) with access to confidential information and/or trade secrets to enter into a nondisclosure and proprietary rights assignment agreement, and (b) each Founder to enter into (i) a one-year noncompetition and non-solicitation agreement, substantially in the form approved by the Board of Directors, and (ii) a non-disclosure and proprietary rights assignment. In addition, the Company shall not amend, modify, terminate, waive, or otherwise alter, in whole or in part, either of the above-referenced agreements or any restricted share agreement between the Company and any employee, without the unanimous consent of the Preferred Directors. Unless otherwise approved by the Board of Directors, all future employees and consultants of the Company who purchase, receive options to purchase, or receive awards of shares is the Company's authorized share capital after the date hereof shall be required to execute restricted shares or option agreements, as applicable, providing for (x) vesting of shares over a four-year period, with the first 25% of such shares vesting following 12

months of continued employment or service, and the remaining shares vesting in equal monthly installments over the following 36 months, and (y) a lock-up agreement provision substantially similar to that in Subsection 2.11. In addition, unless otherwise approved by the Board of Directors, the Company shall retain a "right of first refusal" on employee transfers until the Company's IPO and shall have the right to repurchase unvested shares at cost upon termination of employment of a holder of restricted shares.

5.3 Matters Requiring Investor Director Approval. So long as the holders of Preferred Shares are entitled to elect one or more Preferred Directors, the Company hereby covenants and agrees with each of the Investors that it shall not, and shall procure that its subsidiaries shall not, without approval of the Board of Directors, which approval must include the affirmative vote of all of the Preferred Directors then serving as directors:

(a) make any loan or advance to, or own any shares or other securities of, any subsidiary or other corporation, partnership, or other entity unless it is wholly owned by the Company;

(b) make any loan or advance to any Person, including, without limitation, any employee or director of the Company or any subsidiary, except advances and similar expenditures in the ordinary course of business or under the terms of an employee shares or option plan approved by the Board of Directors;

(c) guarantee, directly or indirectly, any indebtedness except for trade accounts of the Company or any subsidiary arising in the ordinary course of business;

(d) make any investment inconsistent with any investment policy approved by the Board of Directors;

(e) incur any aggregate indebtedness in excess of $750,000 that is not already included in a budget approved by the Board of Directors, other than trade credit incurred in the ordinary course of business;

(f) otherwise enter into or be a party to any transaction with any director, officer, or employee of the Company or its subsidiaries or any "associate" (as defined in Rule 12b-2 promulgated under the Exchange Act) of any such Person, except for (i) transactions made in the ordinary course of business and pursuant to reasonable requirements of the Company's and/or its subsidiaries' business and upon fair and reasonable terms that are approved by a majority of the Board of Directors and (ii) transactions contemplated by the Purchase Agreement;

(g) change the compensation of the Chief Executive Officer of the Company, including approving any option grants or share awards;

(h) change the principal business of the Company or any subsidiary, enter new lines of business, or exit the current line of business;

(i) sell, assign, license, pledge, or encumber material technology or intellectual property, other than licenses granted in the ordinary course of business;

(j) enter into any in-license, asset transfer, merger or acquisition or similar corporate strategic relationship involving Company or subsidiary assets greater than $1,500,000; or

(k) increase the size of the board of directors of the Subsidiary (as defined below).

5.4 Board Matters. Unless otherwise determined by the vote of a majority of the directors then in office, the Board of Directors shall meet at least quarterly in accordance with an agreed-upon schedule. The Company shall reimburse the nonemployee directors for all reasonable out-of-pocket travel expenses incurred (consistent with the Company's travel policy) in connection with attending meetings of the Board of Directors. At each annual or special meeting of shareholders of the Company's wholly owned subsidiary, Kiniksa Pharmaceuticals Corp. (the "**Subsidiary**"), at which an election of directors is held, or pursuant to any written consent of the shareholders of the Subsidiary, the Company shall cause at least two Preferred Directors designated from time to time pursuant to Section 1.2 of that certain Second Amended and Restated Voting Agreement, dated as of the date hereof, by and among the Company and the other parties thereto (as may be amended and/or restated from time to time, the "**Voting Agreement**") as members of the Board of Directors of the Company to be elected to the board of directors of the Subsidiary.

5.5 Successor Indemnification. If the Company or any of its successors or assignees consolidates with or merges into any other Person and is not the continuing or surviving corporation or entity of such consolidation or merger, then to the extent necessary, proper provision shall be made so that the successors and assignees of the Company assume the obligations of the Company with respect to indemnification of members of the Board of Directors as in effect immediately before such transaction, whether such obligations are contained in the Restated Bye-Laws or elsewhere, as the case may be.

5.6 Indemnification Matters. The Company shall enter into indemnification agreements with each Preferred Director (the "**Indemnitees**"). The Company hereby acknowledges that such directors and affiliated funds may have certain rights to indemnification, advancement of expenses and/or insurance provided by one or more of the Investors and certain of their affiliates (collectively, the "**Fund Indemnitors**"), and hereby agrees (a) that it is the indemnitor of first resort (*i.e.*, its obligations to any such Indemnitee are primary and any obligation of the Fund Indemnitors to advance expenses or to provide indemnification for the same expenses or liabilities incurred by such Indemnitee are secondary), (b) that it shall be required to advance the full amount of expenses incurred by such Indemnitee and shall be liable for the full amount of all expenses, judgments, penalties, fines and amounts paid in settlement by or on behalf of any such Indemnitee to the extent legally permitted and as required by the Restated Bye-Laws (or any agreement between the Company and such Indemnitee), without regard to any rights such Indemnitee may have against the Fund Indemnitors, and, (c) that it irrevocably waives, relinquishes and releases the Fund Indemnitors from any and all claims

against the Fund Indemnitors for contribution, subrogation or any other recovery of any kind in respect thereof. The Company further agrees that no advancement or payment by the Fund Indemnitors on behalf of any such Indemnitee with respect to any claim for which such Indemnitee has sought indemnification from the Company shall affect the foregoing and the Fund Indemnitors shall have a right of contribution and/or be subrogated to the extent of such advancement or payment to all of the rights of recovery of such Indemnitee against the Company.

5.7 Right to Conduct Activities. The Company hereby agrees and acknowledges that Baker Bros. Life Sciences, L.P. (together with its Affiliates, "**Baker Bros.**"), HH RSV-XVII Holdings Limited (together with its Affiliates, "**Hillhouse**"), Venrock Healthcare Capital Partners II, L.P. (together with its Affiliates, "**Venrock**"), Sofinnova Venture Partners X, L.P. (together with its Affiliates, "**Sofinnova**") and Deerfield Special Sitautions Fund, L.P. (together with its Affiliates, "**Deerfield**") are professional investment funds, and as such invest in numerous portfolio companies, some of which may be deemed competitive with the Company's business (as currently conducted or as currently proposed to be conducted). The Company hereby agrees that, to the extent permitted under applicable law, Baker Bros., Hillhouse, Venrock, Sofinnova and Deerfield shall not be liable to the Company for any claim arising out of, or based upon, (a) the investment by Baker Bros., Hillhouse, Venrock, Sofinnova or Deerfield in any entity competitive with the Company, or (b) actions taken by any partner, officer or other representative of Baker Bros., Hillhouse, Venrock, Sofinnova or Deerfield to assist any such competitive company, whether or not such action was taken as a member of the board of directors of such competitive company or otherwise, and whether or not such action has a detrimental effect on the Company; provided, however, that the foregoing shall not relieve (i) any of the Investors from liability associated with the unauthorized disclosure of the Company's confidential information obtained pursuant to this Agreement, or (ii) any director or officer of the Company from any liability associated with his or her fiduciary duties to the Company.

5.8 No Publicity. The parties will not, without the prior written consent of the relevant Investor, use in advertising, publicity, marketing communications regarding any Company financing (whether oral or written) or other public communication or filing, the "Baker Brothers", "Hillhouse", "□□", "Gaoling" names or, to its knowledge, the name of any partner, employee, Affiliate and/or controlling person thereof, nor any of their Affiliates and/or controlling persons, nor any trade name, trademark, trade device, service mark, symbol or any abbreviation, or contraction thereof owned by such Investor, except that the Company may make any such disclosure if, upon the advice of counsel, there is no alternative to such disclosure because it is required by applicable law or regulation and the relevant Investors are notified in advance and given reasonable opportunity to minimize such disclosure. In addition, the Company may respond to inquiries about any public disclosure that was required by law or regulation, by confirming the accuracy of such disclosure.

Notwithstanding the foregoing, the Company may disclose the name of the Investors in connection with the provision of any details regarding the agreements executed by the Company and the Investors to any of its executive officers, directors, accountants, counsel and financial advisors, with a need to know such information, provided that such recipient agrees to abide by the foregoing confidentiality obligations.

5.9 Termination of Covenants. The covenants set forth in this Section 5, except for Subsections 5.5 and 5.9, shall terminate and be of no further force or effect (i) immediately before the consummation of the IPO or (ii) upon a Deemed Liquidation Event, as such term is defined in the Restated Bye-Laws, whichever event occurs first.

6. Miscellaneous.

6.1 Successors and Assigns. The rights under this Agreement may be assigned (but only with all related obligations) by a Holder or Founder to a transferee of Registrable Securities or other Common Shares that (a) is an Affiliate of such Holder or Founder, as the case may be, (b) is such Holder's or Founder's Immediate Family Member or trust for the benefit of an individual Holder or Founder or one or more of such Holder's or Founder's Immediate Family Members, or (c) after such transfer, holds at least 10,000 shares of Registrable Securities (subject to appropriate adjustment for share splits, share dividends, combinations, and other recapitalizations); provided, however, that (i) the Company is, within a reasonable time after such transfer, furnished with written notice of the name and address of such transferee and the Registrable Securities or other Common Shares with respect to which such rights are being transferred, and (ii) such transferee agrees in a written instrument delivered to the Company to be bound by and subject to the terms and conditions of this Agreement, including the provisions of Subsection 2.11. For the purposes of determining the number of Registrable Securities or Common Shares held by a transferee, the holdings of a transferee (x) that is an Affiliate or shareholder of a Holder or Founder, (y) who is a Holder's Immediate Family Member, or (z) that is a trust for the benefit of an individual Holder or Founder or such Holder's or Founder's Immediate Family Member shall be aggregated together and with those of the transferring Holder or Founder, as the case may be; provided further that all transferees who would not qualify individually for assignment of rights shall have a single attorney-in-fact for the purpose of exercising any rights, receiving notices, or taking any action under this Agreement. The terms and conditions of this Agreement inure to the benefit of and are binding upon the respective successors and permitted assignees of the parties. Nothing in this Agreement, express or implied, is intended to confer upon any party other than the parties hereto or their respective successors and permitted assignees any rights, remedies, obligations or liabilities under or by reason of this Agreement, except as expressly provided herein.

6.2 Governing Law. This Agreement shall be governed by the internal law of the State of Delaware.

6.3 Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument. Counterparts may be delivered via facsimile, electronic mail (including pdf or any electronic

signature complying with the U.S. federal ESIGN Act of 2000, *e.g.*, www.docusign.com) or other transmission method and any counterpart so delivered shall be deemed to have been duly and validly delivered and be valid and effective for all purposes**.**

6.4 Titles and Subtitles. The titles and subtitles used in this Agreement are for convenience only and are not to be considered in construing or interpreting this Agreement.

6.5 Notices. All notices and other communications given or made pursuant to this Agreement shall be in writing and shall be deemed effectively given upon the earlier of actual receipt or (a) personal delivery to the party to be notified, (b) when sent, if sent by electronic mail or facsimile during the recipient's normal business hours, and if not sent during normal business hours, then on the recipient's next business day, (c) five days after having been sent by registered or certified mail, return receipt requested, postage prepaid, or (d) one business day after the business day of deposit with a nationally recognized overnight courier, freight prepaid, specifying next-day delivery, with written verification of receipt. All communications shall be sent to the respective parties at their addresses as set forth on Schedule A or Schedule B (as applicable) hereto, or to the principal office of the Company and to the attention of the Chief Executive Officer, in the case of the Company, or to such email address, facsimile number, or address as subsequently modified by written notice given in accordance with this Subsection 6.5. Notwithstanding the foregoing, any communications to HH RSV-XVII Holdings Limited shall be sent to Hillhouse Capital Management, Suite 1606, One Exchange Square, 8 Connaught Place, Central, Hong Kong with an email copy to legal@hillhousecap.com. If notice is given to the Company, a copy shall also be sent to Latham & Watkins LLP, 200 Clarendon Street, 27th Floor, Boston, MA 02116, Attention: Johan Brigham.

6.6 Amendments and Waivers. Any term of this Agreement may be amended and the observance of any term of this Agreement may be waived (either generally or in a particular instance, and either retroactively or prospectively) only with the written consent of the Company and the holders of a majority of the Registrable Securities then outstanding; provided that the Company may in its sole discretion waive compliance with Subsection 2.12(c) (and the Company's failure to object promptly in writing after notification of a proposed assignment allegedly in violation of Subsection 2.12(c) shall be deemed to be a waiver); and provided further that any provision hereof may be waived by any waiving party on such party's own behalf, without the consent of any other party. Notwithstanding the foregoing, this Agreement may not be amended or terminated and the observance of any term hereof may not be waived with respect to any Holder without the written consent of such Holder, unless such amendment, termination, or waiver applies to all Holders in the same fashion (it being agreed that a waiver of the provisions of Section 4 with respect to a particular transaction shall be deemed to apply to all Investors in the same fashion if such waiver does so by its terms, notwithstanding the fact that certain Investors may nonetheless, by agreement with the Company, purchase securities in such transaction). Further, this Agreement may not be amended, and no provision hereof may be waived, in each case, in any way which would adversely affect the rights of the Founders hereunder in a manner disproportionately to any adverse effect such amendment or waiver would have on the rights of the Investors hereunder, without also the written consent of the holders of a majority of the Common Shares held by the Founders who are then providing services to the Company or any of its subsidiaries as officers, directors, employees or consultants. The Company shall give prompt notice of any amendment or termination hereof or waiver hereunder to any party hereto that did not consent in writing to such amendment, termination, or waiver. Any amendment, termination, or waiver effected in accordance with this Subsection 6.6 shall be binding on all parties hereto, regardless of whether any such party has consented thereto. No waivers of or exceptions to any term, condition, or provision of this Agreement, in any one or more instances, shall be deemed to be or construed as a further or continuing waiver of any such term, condition, or provision.

6.7 Severability. In case any one or more of the provisions contained in this Agreement is for any reason held to be invalid, illegal or unenforceable in any respect, such invalidity, illegality, or unenforceability shall not affect any other provision of this Agreement, and such invalid, illegal, or unenforceable provision shall be reformed and construed so that it will be valid, legal, and enforceable to the maximum extent permitted by law.

6.8 Aggregation of Shares. All Registrable Securities held or acquired by Affiliates shall be aggregated together for the purpose of determining the availability of any rights under this Agreement and such Affiliated persons may apportion such rights as among themselves in any manner they deem appropriate.

6.9 Additional Investors. Notwithstanding anything to the contrary contained herein, if the Company issues additional Preferred Shares in compliance with the limitations set forth in the Restated Bye-Laws after the date hereof, whether pursuant to the Purchase Agreement or otherwise, any purchaser of such Preferred Shares may become a party to this Agreement by executing and delivering an additional counterpart signature page to this Agreement, and thereafter shall be deemed an "Investor" for all purposes hereunder. No action or consent by the Investors shall be required for such joinder to this Agreement by such additional Investor, so long as such additional Investor has agreed in writing to be bound by all of the obligations as an "Investor" hereunder.

6.10 Entire Agreement; Effect on Prior Agreement. This Agreement (including any Schedules) constitutes the full and entire understanding and agreement among the parties with respect to the subject matter hereof, and any other written or oral agreement relating to the subject matter hereof existing between the parties is expressly canceled. Upon the execution and delivery of this Agreement by the Company and the other parties required to amend the Prior Agreement pursuant to its terms, the Prior Agreement automatically shall terminate and be of no further force and effect and shall be amended and restated in its entirety as set forth in this Agreement.

6.11 Dispute Resolution. The parties (a) hereby irrevocably and unconditionally submit to the jurisdiction of the state courts of New York and to the jurisdiction of the United States District Court for the Southern District of New York for the purpose of any suit, action or other proceeding arising out of or based upon this Agreement, (b) agree not to commence any suit, action or other proceeding arising out of or based upon this Agreement except in the state courts of New York or the United States District Court for the Southern District of New York, and (c) hereby waive, and agree not to assert, by way of motion, as a defense, or otherwise, in any such suit, action or proceeding, any claim that it is not subject personally to the jurisdiction of the above-named courts, that its property is exempt or immune from attachment or execution, that the suit, action or proceeding is brought in an inconvenient forum, that the venue of the suit, action or proceeding is improper or that this Agreement or the subject matter hereof may not be enforced in or by such court.

WAIVER OF JURY TRIAL: EACH PARTY HEREBY WAIVES ITS RIGHTS TO A JURY TRIAL OF ANY CLAIM OR CAUSE OF ACTION BASED UPON OR ARISING OUT OF THIS AGREEMENT, THE OTHER TRANSACTION DOCUMENTS, THE SECURITIES OR

THE SUBJECT MATTER HEREOF OR THEREOF. THE SCOPE OF THIS WAIVER IS INTENDED TO BE ALL-ENCOMPASSING OF ANY AND ALL DISPUTES THAT MAY BE FILED IN ANY COURT AND THAT RELATE TO THE SUBJECT MATTER OF THIS TRANSACTION, INCLUDING, WITHOUT LIMITATION, CONTRACT CLAIMS, TORT CLAIMS (INCLUDING NEGLIGENCE), BREACH OF DUTY CLAIMS, AND ALL OTHER COMMON LAW AND STATUTORY CLAIMS. THIS SECTION HAS BEEN FULLY DISCUSSED BY EACH OF THE PARTIES HERETO AND THESE PROVISIONS WILL NOT BE SUBJECT TO ANY EXCEPTIONS. EACH PARTY HERETO HEREBY FURTHER WARRANTS AND REPRESENTS THAT SUCH PARTY HAS REVIEWED THIS WAIVER WITH ITS LEGAL COUNSEL, AND THAT SUCH PARTY KNOWINGLY AND VOLUNTARILY WAIVES ITS JURY TRIAL RIGHTS FOLLOWING CONSULTATION WITH LEGAL COUNSEL.

6.12 Delays or Omissions. No delay or omission to exercise any right, power, or remedy accruing to any party under this Agreement, upon any breach or default of any other party under this Agreement, shall impair any such right, power, or remedy of such nonbreaching or nondefaulting party, nor shall it be construed to be a waiver of or acquiescence to any such breach or default, or to any similar breach or default thereafter occurring, nor shall any waiver of any single breach or default be deemed a waiver of any other breach or default theretofore or thereafter occurring. All remedies, whether under this Agreement or by law or otherwise afforded to any party, shall be cumulative and not alternative.

6.13 Acknowledgment. The Company acknowledges that the Investors are in the business of venture capital investing and therefore review the business plans and related proprietary information of many enterprises, including enterprises which may have products or services which compete directly or indirectly with those of the Company. Nothing in this Agreement shall preclude or in any way restrict the Investors from investing or participating in any particular enterprise whether or not such enterprise has products or services which compete with those of the Company; provided, however, that the foregoing shall not relieve (i) any of the Investors from liability associated with the unauthorized disclosure of the Company's confidential information obtained pursuant to this Agreement, or (ii) any director or officer of the Company from any liability associated with his or her fiduciary duties to the Company.

[*Remainder of Page Intentionally Left Blank*]

IN WITNESS WHEREOF, the parties have executed this Agreement as of the date first written above.

KINIKSA PHARMACEUTICALS, LTD.

By: /s/ Sanj K. Patel
Name: Sanj K. Patel
Title: Chief Executive Officer

INVESTORS:

667, L.P.

BY: **BAKER BROS. ADVISORS LP**, management company and investment adviser to **667, L.P.**, pursuant to authority granted to it by Baker Biotech Capital, L.P., general partner to 667, L.P., and not as the general partner.

By: /s/ Scott Lessing
Scott Lessing
President

BAKER BROTHERS LIFE SCIENCES, L.P.

By: **BAKER BROS. ADVISORS LP,**
management company and investment adviser to **Baker Brothers Life Sciences, L.P**., pursuant to authority granted to it by Baker Brothers Life Sciences Capital, L.P., general partner to Baker Brothers Life Sciences, L.P., and not as the general partner.

By: /s/ Scott Lessing
Scott Lessing
President

INVESTORS:

MERIDIAN SMALL CAP GROWTH FUND

By: its Investment Adviser
ArrowMark Partners

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

ARROWMARK FUNDAMENTAL OPPORTUNITY FUND, L.P.

By: its General Partner
ArrowMark Partners GP, LLC

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

LOOKFAR INVESTMENTS LLC

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

IRON HORSE INVESTMENTS LLC

By: its Investment Adviser
ArrowMark Partners

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

THB IRON ROSE LLC

By: its Investment Adviser
ArrowMark Partners

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

THB IRON ROSE LLC, LIFE SCIENCE PORTFOLIO

By: its Investment Adviser
ArrowMark Partners

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

/s/ Tony Yao
Tony Yao

MERIDIAN GROWTH FUND

By: its Investment Adviser
ArrowMark Partners

By: /s/ David Corkins
Name: David Corkins
Title: Managing Member

INVESTORS:

EEJ, LLC

By: /s/ Edward F. Keely
Name: Edward F. Keely
Title: Manager

INVESTORS:

HH RSV-XVII HOLDINGS LIMITED

By: /s/ Colm O'Connell
Name: Colm O'Connell
Title: Authorized signatory

INVESTORS:

MOSSROCK CAPITAL, LLC

By: /s/ Thomas Malley
Name: Thomas Malley
Title: President

INVESTORS:

NEW DIRECTION IRA, INC. FBO ROBERT DESNICK ROTH IRA

By: /s/ Robert Desnick
Name: Robert Desnick
Title: Principal

Address: 170 E. 93rd St.
New York, NY 10128

INVESTORS:

VENROCK HEALTHCARE CAPITAL PARTNERS II, L.P
BY: VHCP MANAGEMENT II, LLC
ITS: GENERAL PARTNER

By: /s/ David L. Stepp
Name: David L. Stepp
Title: Authorized Signature

Address:

3340 Hillview Avenue
Palo Alto, CA 94304

VHCP CO-INVESTMENT HOLDINGS II, LLC
BY: VHCP MANAGEMENT II, LLC
ITS: MANAGER

By: /s/ David L. Stepp
Name: David L. Stepp
Title: Authorized Signature

Address:

3340 Hillview Avenue
Palo Alto, CA 94304

INVESTORS:

STURM FAMILY CAPITAL, LLLP

By: /s/ Donald L. Sturm
Name: Donald L. Sturm, Managing Partner
Title:

Address:
3033 E. 1st Avenue, Suite 300
Denver, CO 80206

INVESTORS:

BOXER CAPITAL LLC

By: /s/ Aaron Davis
Name: Aaron Davis
Title: CEO

Address:

11682 El Camino Real, Suite 320
San Diego, CA, 92130

INVESTORS:

FORESITE CAPITAL FUND IV, L.P.
By: **FORESITE CAPITAL MANAGEMENT IV, LLC**
Its: **GENERAL PARTNER**

By: /s/ Dennis D. Ryan
Name: Dennis D. Ryan
Title: Chief Financial Officer

Address:

600 Montgomery Street, Suite 4500
San Francisco, CA 94111

INVESTORS:

CORMORANT PRIVATE HEALTHCARE FUND I, L.P.

By: /s/ Bihua Chen
Name: Bihua Chen
Title: Managing Member of the GP

Address:

200 Clarendon Street, 52nd Floor
Boston, MA 02116

CORMORANT GLOBAL HEALTHCARE MASTER FUND, LP

By: /s/ Bihua Chen
Name: Bihua Chen
Title: Managing Member of the GP

Address:

200 Clarendon Street, 52nd Floor
Boston, MA 02116

CRMA SPV, L.P.

By: /s/ Bihua Chen
Name: Bihua Chen
Title: CEO/CIO of Cormorant Asset Management LLC
Its: Attorney-In-Fact

Address:

PO Box 309
Ugland House
Grand Cayman
KY1-1104 Cayman Islands

INVESTORS:

SOFINNOVA VENTURE PARTNERS X, L.P.

By: **SOFINNOVA MANAGEMENT X, L.L.C.**
Its: **GENERAL PARTNER**

By: /s/ Anand Mehra
Name: Anand Mehra
Title: Managing Member

Address:

3000 Sand Hill Road, Bldg. 4, Suite 250
Menlo Park, CA 94025

SIGNATURE PAGE TO AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

INVESTORS:

DEERFIELD SPECIAL SITUATIONS FUND, L.P.
BY: DEERFIELD MGMT, L.P., GENERAL PARTNER
BY: J.E. FLYNN CAPITAL, LLC, GENERAL PARTNER

By: /s/ David J. Clark
Name: David J. Clark
Title: Authorized Signatory

Address:

780 Third Avenue
New York, NY 10017

SIGNATURE PAGE TO AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

INVESTORS:

/s/ Stephen Frank Mahoney
Stephen Frank Mahoney

Address:

[xxxx]

SIGNATURE PAGE TO AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

INVESTORS:

/s/ Sanj J. Patel
Sanj K. Patel

Address:

[xxxx]

SIGNATURE PAGE TO AMENDED AND RESTATED INVESTORS' RIGHTS AGREEMENT

INVESTORS:

/s/ Thomas Beetham
Thomas W. Beetham

Address:

[xxxx]

INVESTORS:

/s/ Christopher Heberlig
Christopher Heberlig

Address:

[xxxx]

INVESTORS:

/s/ Carsten Boess
Carsten Boess

Address:

[xxxx]

INVESTORS:

/s/ Rasmus Holm-Jorgensen
Rasmus Holm-Jorgensen

Address:

[xxxx]

INVESTORS:

/s/ Aaron Isadore Young
Aaron Isadore Young

Address:

[xxxx]

SCHEDULE A

Investors

667, L.P.
667 Madison Avenue, 21st Floor
New York, New York 10065

Baker Brothers Life Sciences, L.P.
667 Madison Avenue, 21st Floor
New York, New York 10065

New Direction IRA, Inc.
FBO Robert Desnick Roth IRA
1070 W. Century Drive Suite 101
Louisville, CO 80027

Sanj K. Patel
[xxxx]

Stephen Frank Mahoney
[xxxx]

Thomas W. Beetham
[xxxx]

Christopher Heberlig
[xxxx]

Carsten Boess
[xxxx]

Rasmus Holm-Jorgensen
[xxxx]

Gregory Alex Grabowski
[xxxx]

Aaron Isadore Young
[xxxx]

Eben P. Tessari
[xxxx]

Meridian Small Cap Growth Fund
100 Fillmore Street, Suite 325
Denver, CO 80206

Arrowpoint Fundamental Opportunity Fund, L.P.
100 Fillmore Street, Suite 325
Denver, CO 80206

Lookfar Investments LLC
100 Fillmore Street, Suite 325
Denver, CO 80206

Iron Horse Investments LLC
100 Fillmore Street, Suite 325
Denver, CO 80206

THB Iron Rose LLC
100 Fillmore Street, Suite 325
Denver, CO 80206

THB Iron Rose LLC, Life Science Portfolio
100 Fillmore Street, Suite 325
Denver, CO 80206

Tony Yao
[xxxx]

EEJ, LLC
3033 E. 1st Avenue, Suite 415
Denver, CO 80206

HH RSV-XVII Holdings Limited
Hillhouse Capital Management, Suite 1606
One Exchange Square
8 Connaught Place
Central, Hong Kong

Mossrock Capital, LLC
19 Martin Lane
Englewood CO 80113

Edward Schuchman, Ph.D, as Trustee for the
Desnick / Herzig 2012 GST Trust UAD 10/23/12
[xxxx]

Venrock Healthcare Capital Partners II, L.P
3340 Hillview Avenue
Palo Alto, CA 94304

VHCP Co-Investment Holdings II, LLC
3340 Hillview Avenue
Palo Alto, CA 94304

Sturm Family Capital, LLLP
3033 E. 1st Avenue, Suite 300
Denver, CO 80206

Boxer Capital LLC
Attn: Christopher Fuglesang
11682 El Camino Real, Suite 320
San Diego, CA, 92130

Foresite Capital Fund IV, L.P.
600 Montgomery Street, Suite 4500
San Francisco, CA 94111

Cormorant Private Healthcare Fund I, L.P.
200 Clarendon Street, 52nd Floor
Boston, MA 02116

Cormorant Global Healthcare Master Fund, LP
200 Clarendon Street, 52nd Floor
Boston, MA 02116

CRMA SPV, L.P.
PO Box 309
Ugland House
Grand Cayman
KY1-1104 Cayman Islands

Soffinova Venture Partners X, L.P.
3000 Sand Hill Road, Bldg. 4, Suite 250
Menlo Park, CA 94025

Deerfield Special Situations Fund, L.P.
780 Third Avenue
New York, NY 10017

SCHEDULE B

Founders

Name and Address

Sanj K. Patel
[xxxx]

Sanjiv K. Patel, as Trustee
for the Manisha S. Patel
2016 Irrevocable Trust
[xxxx]

Stephen Frank Mahoney
[xxxx]

Krisha S. Mahoney as Trustee
for the Stephen F. Mahoney
2016 Irrevocable Trust
[xxxx]

Thomas W. Beetham
[xxxx]

Christopher Heberlig
[xxxx]

Carsten Boess
[xxxx]

Rasmus Holm-Jorgensen
[xxxx]

Gregory Alex Grabowski
[xxxx]

Aaron Isadore Young
[xxxx]

Eben P. Tessari
[xxxx]

Jennifer Lynne Mason
[xxxx]

Mickenzie Elizabeth Gallagher
[xxxx]

EXHIBIT 10.1

PLAN AMENDMENT

Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company (the "Company"), has adopted the 2015 Equity Incentive Plan (the "Plan"). Unless otherwise defined herein, all capitalized terms shall have the meaning set forth in the Plan.

Section 3, Paragraph (a) of the Plan shall be amended in its entirety to read as follows:

"(a) The number of Shares which may be issued from time to time pursuant to this Plan shall be thirteen million ninety nine thousand six hundred and fourteen (13,099,614), or the equivalent of such number of Shares after the Administrator, in its sole discretion, has interpreted the effect of any share split, share dividend, combination, recapitalization or similar transaction in accordance with Paragraph 24 of the Plan."

Except as expressly amended hereby, the Plan shall remain unchanged and in full force and effect and is hereby ratified and confirmed.

Adopted by the Company's Board of Directors: March 6, 2017

Adopted by the Company's Shareholders: March 6, 2017

IN WITNESS WHEREOF, the undersigned has acknowledged this Plan Amendment this 8th day of March, 2017.

By: /s/ Sanj K. Patel
Sanj K. Patel
President and Chief Executive Officer

SIGNATURE PAGE TO PLAN AMENDMENT

KINIKSA PHARMACEUTICALS, LTD.

2015 EQUITY INCENTIVE PLAN

1. DEFINITIONS.

Unless otherwise specified or unless the context otherwise requires, the following terms, as used in this Kiniksa Pharmaceuticals, Ltd. 2015 Equity Incentive Plan, have the following meanings:

Administrator means the Board of Directors, unless it has delegated power to act on its behalf to the Committee, in which case the Administrator means the Committee.

Affiliate means a corporation which, for purposes of Section 424 of the Code, is a parent or subsidiary of the Company, direct or indirect.

Agreement means an agreement between the Company and a Participant delivered pursuant to the Plan and pertaining to a Share Right, in such form as the Administrator shall approve.

Board of Directors means the Board of Directors of the Company.

California Participant means a Participant who resides in the State of California.

Cause means, with respect to a Participant (a) dishonesty with respect to the Company or any Affiliate, (b) insubordination, substantial malfeasance or non-feasance 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 Affiliate, and (e) conduct substantially prejudicial to the business of the Company or any Affiliate; provided, however, that any provision in an agreement between a Participant and the Company or an Affiliate, 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.

Code means the United States Internal Revenue Code of 1986, as amended including any successor statute, regulation and guidance thereto.

Committee means the committee of the Board of Directors to which the Board of Directors has delegated power to act under or pursuant to the provisions of the Plan.

Common Shares means Class A common shares of the Company, $0.0001 par value per share.

Company means Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

Consultant means any natural person who is an advisor or consultant that provides bona fide services to the Company or its Affiliates, provided that such services are not in connection with the offer or sale of securities in a capital raising transaction, and do not directly or indirectly promote or maintain a market for the Company's or its Affiliates' securities.

Disability or Disabled means permanent and total disability as defined in Section 22(e)(3) of the Code.

Employee means any employee of the Company or of an Affiliate (including, without limitation, an employee who is also serving as an officer or director of the Company or of an Affiliate), designated by the Administrator to be eligible to be granted one or more Share Rights under the Plan.

Exchange Act means the Securities Exchange Act of 1934, as amended.

Fair Market Value of a Share of Common Shares means:

(1) If the Common Shares are listed on a national securities exchange or traded in the over-the-counter market and sales prices are regularly reported for the Common Shares, the closing or, if not applicable, the last price of the Common Shares on the composite tape or other comparable reporting system for the trading day on the applicable date and if such applicable date is not a trading day, the last market trading day prior to such date;

(2) If the Common Shares are not traded on a national securities exchange but are traded on the over-the-counter market, if sales prices are not regularly reported for the Common Shares for the trading day referred to in clause (1), and if bid and asked prices for the Common Shares are regularly reported, the mean between the bid and the asked price for the Common Shares at the close of trading in the over-the-counter market for the trading day on which Common Shares was traded on the applicable date and if such applicable date is not a trading day, the last market trading day prior to such date; and

(3) If the Common Shares are neither listed on a national securities exchange nor traded in the over-the-counter market, such value as the Administrator, in good faith, shall determine.

ISO means an option intended to qualify as an incentive stock option under Section 422 of the Code.

Non-Qualified Option means an option which is not intended to qualify as an ISO.

Option means an ISO or Non-Qualified Option granted under the Plan.

Participant means an Employee, director or Consultant of the Company or an Affiliate to whom one or more Share Rights are granted under the Plan. As used herein, "Participant" shall include "Participant's Survivors" where the context requires.

Plan means this Kiniksa Pharmaceuticals, Ltd. 2015 Equity Incentive Plan, as may be amended and/or restated from time to time.

Securities Act means the Securities Act of 1933, as amended.

Shares means shares of the Common Shares as to which Share Rights have been or may be granted under the Plan or any shares into which the Shares are changed or for which they are exchanged within the provisions of Paragraph 3 of the Plan. The Shares issued under the Plan may be authorized and unissued shares or shares held by the Company in its treasury, or both.

Share-Based Award means a grant by the Company under the Plan of an equity award or an equity based award which is not an Option or a Share Grant.

Share Grant means a grant by the Company of Shares under the Plan.

Share Right means a right to Shares or the value of Shares of the Company granted pursuant to the Plan — an ISO, a Non-Qualified Option, a Share Grant or a Share-Based Award.

Survivor means a deceased Participant's legal representatives and/or any person or persons who acquired the Participant's rights to a Share Right by will or by the laws of descent and distribution.

2. PURPOSES OF THE PLAN.

The Plan is intended to encourage ownership of Shares by Employees and directors of and certain Consultants to the Company and its Affiliates in order to attract and retain such people, to induce them to work for the benefit of the Company or of an Affiliate and to provide additional incentive for them to promote the success of the Company or of an Affiliate. The Plan provides for the granting of ISOs, Non-Qualified Options, Share Grants and Share-Based Awards.

3. SHARES SUBJECT TO THE PLAN.

(a) The number of Shares which may be issued from time to time pursuant to this Plan shall be ( ), or the equivalent of such number of Shares after the Administrator, in its sole discretion, has interpreted the effect of any share split, share dividend, combination, recapitalization or similar transaction in accordance with Paragraph 24 of the Plan.

(b) If an Option ceases to be "outstanding", in whole or in part (other than by exercise), or if the Company shall reacquire (at not more than its original issuance price) any Shares issued pursuant to a Share Grant or Share-Based Award, or if any Share Right expires or

is forfeited, cancelled, or otherwise terminated or results in any Shares not being issued, the unissued or reacquired Shares which were subject to such Share Right shall again be available for issuance from time to time pursuant to this Plan. Notwithstanding the foregoing, if a Share Right is exercised, in whole or in part, by tender of Shares or if the Company or an Affiliate's tax withholding obligation is satisfied by withholding Shares, the number of Shares deemed to have been issued under the Plan for purposes of the limitation set forth in Paragraph 3(a) above shall be the number of Shares that were subject to the Share Right or portion thereof, and not the net number of Shares actually issued. However, in the case of ISOs, the foregoing provisions shall be subject to any limitations under the Code.

4. ADMINISTRATION OF THE PLAN.

The Administrator of the Plan will be the Board of Directors, except to the extent the Board of Directors delegates its authority to the Committee, in which case the Committee shall be the Administrator. Subject to the provisions of the Plan, the Administrator is authorized to:

(a) Interpret the provisions of the Plan and all Share Rights and to make all rules and determinations which it deems necessary or advisable for the administration of the Plan;

(b) Determine which Employees, directors and Consultants shall be granted Share Rights;

(c) Determine the number of Shares for which a Share Right or Share Rights shall be granted;

(d) Specify the terms and conditions upon which a Share Right or Share Rights may be granted;

(e) Amend any term or condition of any outstanding Share Right, including, without limitation, to reduce or increase the exercise price or purchase price, accelerate the vesting schedule or extend the expiration date, provided that (i) such term or condition as amended is permitted by the Plan; (ii) any such amendment shall not impair the rights of a Participant under any Share Right previously granted without such Participant's consent or in the event of death of the Participant the Participant's Survivors; and (iii) any such amendment shall be made only after the Administrator determines whether such amendment would cause any adverse tax consequences to the Participant, including, but not limited to, the annual vesting limitation contained in Section 422(d) of the Code and described in Paragraph 6(b)(iv) below with respect to ISOs and pursuant to Section 409A of the Code;

(f) Buy out for a payment in cash or Shares, a Share Right previously granted and/or cancel any such Share Right and grant in substitution therefor other Share Rights, covering the same or a different number of Shares and having an exercise price or purchase price per share which may be lower or higher than the exercise price or purchase price of the cancelled Share Right, based on such terms and conditions as the Administrator shall establish and the Participant shall accept; and

(g) Adopt any sub-plans applicable to residents of any specified jurisdiction as it deems necessary or appropriate in order to comply with or take advantage of any tax or other

laws applicable to the Company, any Affiliate or to Participants or to otherwise facilitate the administration of the Plan, which sub-plans may include additional restrictions or conditions applicable to Share Rights or Shares issuable pursuant to a Share Right;

provided, however, that all such interpretations, rules, determinations, terms and conditions shall be made and prescribed in the context of not causing any adverse tax consequences under Section 409A of the Code and preserving the tax status under Section 422 of the Code of those Options which are designated as ISOs. Subject to the foregoing, the interpretation and construction by the Administrator of any provisions of the Plan or of any Share Right granted under it shall be final, unless otherwise determined by the Board of Directors, if the Administrator is the Committee. In addition, if the Administrator is the Committee, the Board of Directors may take any action under the Plan that would otherwise be the responsibility of the Committee.

To the extent permitted under applicable law, the Board of Directors or the Committee may allocate all or any portion of its responsibilities and powers to any one or more of its members and may delegate all or any portion of its responsibilities and powers to any other person selected by it. The Board of Directors or the Committee may revoke any such allocation or delegation at any time.

5. ELIGIBILITY FOR PARTICIPATION.

The Administrator will, in its sole discretion, name the Participants in the Plan; provided, however, that each Participant must be an Employee, director or Consultant of the Company or of an Affiliate at the time a Share Right is granted. Notwithstanding the foregoing, the Administrator may authorize the grant of a Share Right to a person not then an Employee, director or Consultant of the Company or of an Affiliate; provided, however, that the actual grant of such Share Right shall be conditioned upon such person becoming eligible to become a Participant at or prior to the time of the execution of the Agreement evidencing such Share Right. ISOs may be granted only to Employees who are deemed to be residents of the United States for tax purposes. Non-Qualified Options, Share Grants and Share-Based Awards may be granted to any Employee, director or Consultant of the Company or an Affiliate. The granting of any Share Right to any individual shall neither entitle that individual to, nor disqualify him or her from, participation in any other grant of Share Rights or any grant under any other benefit plan established by the Company or any Affiliate for Employees, directors or Consultants.

6. TERMS AND CONDITIONS OF OPTIONS.

Each Option shall be set forth in writing in an Option Agreement, duly executed by the Company and, to the extent required by law or requested by the Company, by the Participant. The Administrator may provide that Options be granted subject to such terms and conditions, consistent with the terms and conditions specifically required under this Plan, as the Administrator may deem appropriate including, without limitation, subsequent approval by the shareholders of the Company of this Plan or any amendments thereto. The Option Agreements shall be subject to at least the following terms and conditions:

(a) Non-Qualified Options: Each Option intended to be a Non-Qualified Option shall be subject to the terms and conditions which the Administrator determines to be appropriate and in the best interest of the Company, subject to the following minimum standards for any such Non-Qualified Option:

(i) Exercise Price: Each Option Agreement shall state the exercise price (per share) of the Shares covered by each Option, which exercise price shall be determined by the Administrator and shall be at least equal to the Fair Market Value per share of Common Shares on the date of grant of the Option, provided that if the exercise price is less than Fair Market Value, the terms of such Option must comply with the requirements of Section 409A of the Code unless granted to a Consultant to whom Section 409A of the Code does not apply.

(ii) Number of Shares: Each Option Agreement shall state the number of Shares to which it pertains.

(iii) Option Periods: Each Option Agreement shall state the date or dates on which it first is exercisable and the date after which it may no longer be exercised, and may provide that the Option rights accrue or become exercisable in installments over a period of months or years, or upon the occurrence of certain conditions or the attainment of stated goals or events. For California Participants, the exercise period of the Option set forth in the Option Agreement shall not be more than 120 months from the date of grant.

(iv) Option Conditions: Exercise of any Option may be conditioned upon the Participant's execution of a Share purchase agreement in form satisfactory to the Administrator providing for certain protections for the Company and its other shareholders, including requirements that:

A. The Participant's or the Participant's Survivors' right to sell or transfer the Shares may be restricted; and

B. The Participant or the Participant's Survivors may be required to execute letters of investment intent and must also acknowledge that the Shares will bear legends noting any applicable restrictions.

(v) Term of Option: Each Option shall terminate not more than ten years from the date of the grant or at such earlier time as the Option Agreement may provide.

(b) ISOs: Each Option intended to be an ISO shall be issued only to an Employee who is deemed to be a resident of the United States for tax purposes, and shall be subject to the following terms and conditions, with such additional restrictions or changes as the Administrator determines are appropriate but not in conflict with Section 422 of the Code and relevant regulations and rulings of the Internal Revenue Service:

(i) Minimum standards: The ISO shall meet the minimum standards required of Non-Qualified Options, as described in Paragraph 6(a) above, except clause (i) and (v) thereunder.

(ii) Exercise Price: Immediately before the ISO is granted, if the Participant owns, directly or by reason of the applicable attribution rules in Section 424(d) of the Code:

A. 10% or less of the total combined voting power of all classes of shares of the Company or an Affiliate, the exercise price per share of the Shares covered by each ISO shall not be less than 100% of the Fair Market Value per share of the Common Shares on the date of grant of the Option; or

B. More than 10% of the total combined voting power of all classes of shares of the Company or an Affiliate, the exercise price per share of the Shares covered by each ISO shall not be less than 110% of the Fair Market Value per share of the Common Shares on the date of grant of the Option.

(iii) Term of Option: For Participants who own:

A. 10% or less of the total combined voting power of all classes of shares of the Company or an Affiliate, each ISO shall terminate not more than ten years from the date of the grant or at such earlier time as the Option Agreement may provide; or

B. More than 10% of the total combined voting power of all classes of shares of the Company or an Affiliate, each ISO shall terminate not more than five years from the date of the grant or at such earlier time as the Option Agreement may provide.

(iv) Limitation on Yearly Exercise: The Option Agreements shall restrict the amount of ISOs which may become exercisable in any calendar year (under this or any other ISO plan of the Company or an Affiliate) so that the aggregate Fair Market Value (determined on the date each ISO is granted) of the shares with respect to which ISOs are exercisable for the first time by the Participant in any calendar year does not exceed $100,000.

7. TERMS AND CONDITIONS OF SHARE GRANTS.

Each Share Grant to a Participant shall state the principal terms in an Agreement duly executed by the Company and, to the extent required by law or requested by the Company, by the Participant. For California Participants, each Share Grant shall be issued within ten (10) years from the earlier of the date the Plan is adopted or approved by the Company's shareholders. The Agreement shall be in a form approved by the Administrator and shall contain

terms and conditions which the Administrator determines to be appropriate and in the best interest of the Company, subject to the following minimum standards:

(a) Each Agreement shall state the purchase price per share, if any, of the Shares covered by each Share Grant, which purchase price shall be determined by the Administrator but shall not be less than the minimum consideration required by Bermuda law, if any, on the date of the grant of the Share Grant;

(b) Each Agreement shall state the number of Shares to which the Share Grant pertains; and

(c) Each Agreement shall include the terms of any right of the Company to restrict or reacquire the Shares subject to the Share Grant, including the time and events upon which such rights shall accrue and the purchase price therefor, if any.

8. TERMS AND CONDITIONS OF OTHER SHARE-BASED AWARDS.

The Administrator shall have the right to grant other Share-Based Awards based upon the Common Shares having such terms and conditions as the Administrator may determine, including, without limitation, the grant of Shares based upon certain conditions, the grant of securities convertible into Shares and the grant of share appreciation rights, phantom share awards or share units. The principal terms of each Share-Based Award shall be set forth in an Agreement, duly executed by the Company and, to the extent required by law or requested by the Company, by the Participant. The Agreement shall be in a form approved by the Administrator and shall contain terms and conditions which the Administrator determines to be appropriate and in the best interest of the Company.

The Company intends that the Plan and any Share-Based Awards granted hereunder be exempt from the application of Section 409A of the Code or meet the requirements of paragraphs (2), (3) and (4) of subsection (a) of Section 409A of the Code, to the extent applicable, and be operated in accordance with Section 409A so that any compensation deferred under any Share-Based Award (and applicable investment earnings) shall not be included in income under Section 409A of the Code. Any ambiguities in the Plan shall be construed to effect the intent as described in this Paragraph 8.

9. EXERCISE OF OPTIONS AND ISSUE OF SHARES.

An Option (or any part or installment thereof) shall be exercised by giving written notice to the Company or its designee (in a form acceptable to the Administrator, which may include electronic notice), together with provision for payment of the aggregate exercise price in accordance with this Paragraph for the Shares as to which the Option is being exercised, and upon compliance with any other condition(s) set forth in the Option Agreement. Such notice shall be signed by the person exercising the Option (which signature may be provided electronically in a form acceptable to the Administrator), shall state the number of Shares with respect to which the Option is being exercised and shall contain any representation required by the Plan or the Option Agreement. Payment of the exercise price for the Shares as to which such Option is being exercised shall be made (a) in United States dollars in cash or by check, or (b) at the discretion of the Administrator, through delivery of Common Shares held for at least six

months (if required to avoid negative accounting treatment) having a Fair Market Value equal as of the date of the exercise to the aggregate cash exercise price for the number of Shares as to which the Option is being exercised, or (c) at the discretion of the Administrator, by having the Company retain from the Shares otherwise issuable upon exercise of the Option, a number of Shares having a Fair Market Value equal as of the date of exercise to the aggregate exercise price for the number of Shares as to which the Option is being exercised, or (d) at the discretion of the Administrator (after consideration of applicable securities, tax and accounting implications), by delivery of the grantee's personal recourse note bearing interest payable not less than annually at no less than 100% of the applicable Federal rate, as defined in Section 1274(d) of the Code, or (e) at the discretion of the Administrator, in accordance with a cashless exercise program established with a securities brokerage firm, and approved by the Administrator, or (f) at the discretion of the Administrator, by any combination of (a), (b), (c), (d) and (e) above or (g) at the discretion of the Administrator, by payment of such other lawful consideration as the Administrator may determine. Notwithstanding the foregoing, the Administrator shall accept only such payment on exercise of an ISO as is permitted by Section 422 of the Code.

The Company shall then reasonably promptly deliver the Shares as to which such Option was exercised to the Participant (or to the Participant's Survivors, as the case may be). In determining what constitutes "reasonably promptly," it is expressly understood that the issuance and delivery of the Shares may be delayed by the Company in order to comply with any law or regulation (including, without limitation, state securities or "blue sky" laws) which requires the Company to take any action with respect to the Shares prior to their issuance. The Shares shall, upon delivery, be fully paid, non-assessable Shares.

10. PAYMENT IN CONNECTION WITH THE ISSUANCE OF SHARE GRANTS AND SHARE-BASED AWARDS AND ISSUE OF SHARES.

Any Share Grant or Share-Based Award requiring payment of a purchase price for the Shares as to which such Share Grant or Share-Based Award is being granted shall be made (a) in United States dollars in cash or by check, or (b) at the discretion of the Administrator, through delivery of Common Shares held for at least six months (if required to avoid negative accounting treatment) and having a Fair Market Value equal as of the date of payment to the purchase price of the Share Grant or Share-Based Award, or (c) at the discretion of the Administrator (after consideration of applicable securities, tax and accounting implications), by delivery of the grantee's personal recourse note bearing interest payable not less than annually at no less than 100% of the applicable Federal rate, as defined in Section 1274(d) of the Code, or (d) at the discretion of the Administrator, by any combination of (a), (b) and (c) above, or (e) at the discretion of the Administrator, by payment of such other lawful consideration as the Administrator may determine.

The Company shall when required by the applicable Agreement, reasonably promptly deliver the Shares as to which such Share Grant or Share-Based Award was made to the Participant (or to the Participant's Survivors, as the case may be), subject to any escrow provision set forth in the applicable Agreement. In determining what constitutes "reasonably promptly," it is expressly understood that the issuance and delivery of the Shares may be delayed by the Company in order to comply with any law or regulation (including, without limitation,

state securities or "blue sky" laws) which requires the Company to take any action with respect to the Shares prior to their issuance.

11. RIGHTS AS A SHAREHOLDER.

No Participant to whom a Share Right has been granted shall have rights as a shareholder with respect to any Shares covered by such Share Right except after due exercise of an Option or issuance of Shares as set forth in any Agreement, tender of the aggregate exercise or purchase price, if any, for the Shares being purchased and registration of the Shares in the Company's share register in the name of the Participant.

12. ASSIGNABILITY AND TRANSFERABILITY OF SHARE RIGHTS.

By its terms, a Share Right granted to a Participant shall not be transferable by the Participant other than (i) by will or by the laws of descent and distribution, or (ii) as approved by the Administrator in its discretion and set forth in the applicable Agreement provided that no Share Right may be transferred by a Participant for value. For California Participants, Share Rights shall not be transferable by the Participant other than by will or by the laws of descent and distribution, to a revocable trust, or as permitted by Rule 701 of the Securities Act. Notwithstanding the foregoing, an ISO transferred except in compliance with clause (i) above shall no longer qualify as an ISO. The designation of a beneficiary of a Share Right by a Participant, with the prior approval of the Administrator and in such form as the Administrator shall prescribe, shall not be deemed a transfer prohibited by this Paragraph. Except as provided above during the Participant's lifetime a Share Right shall only be exercisable by or issued to such Participant (or his or her legal representative) and shall not be assigned, pledged or hypothecated in any way (whether by operation of law or otherwise) and shall not be subject to execution, attachment or similar process. Any attempted transfer, assignment, pledge, hypothecation or other disposition of any Share Right or of any rights granted thereunder contrary to the provisions of this Plan, or the levy of any attachment or similar process upon a Share Right, shall be null and void.

13. EFFECT ON OPTIONS OF TERMINATION OF SERVICE OTHER THAN FOR CAUSE OR DEATH OR DISABILITY.

Except as otherwise provided in a Participant's Option Agreement, in the event of a termination of service (whether as an Employee, director or Consultant) with the Company or an Affiliate before the Participant has exercised an Option, the following rules apply:

(a) A Participant who ceases to be an Employee, director or Consultant of the Company or of an Affiliate for any reason other than termination for Cause, Disability, or death, for which events there are special rules in Paragraphs 14, 15, and 16, respectively, may exercise any Option granted to him or her to the extent that the Option is exercisable on the date of such termination of service, but only within such term as the Administrator has designated in a Participant's Option Agreement.

(b) Except as provided in Subparagraph (c) below, or Paragraph 15 or 16, in no event may an Option intended to be an ISO, be exercised later than three months after the Participant's termination of employment. For Options granted to California Participants, an Option must be

exercisable for at least thirty (30) days from the date of a Participant's termination of employment.

(c) The provisions of this Paragraph, and not the provisions of Paragraph 15 or 16, shall apply to a Participant who subsequently becomes Disabled or dies after the termination of employment, director status or consultancy; provided, however, in the case of a Participant's Disability or death within three months after the termination of employment, director status or consultancy, the Participant or the Participant's Survivors may exercise the Option within one year after the date of the Participant's termination of service, but in no event after the date of expiration of the term of the Option.

(d) Notwithstanding anything herein to the contrary, if subsequent to a Participant's termination of employment, termination of director status or termination of consultancy, but prior to the exercise of an Option, the Administrator determines that, either prior or subsequent to the Participant's termination, the Participant engaged in conduct which would constitute Cause, then such Participant shall forthwith cease to have any right to exercise any Option.

(e) A Participant to whom an Option has been granted under the Plan who is absent from the Company or an Affiliate because of temporary disability (any disability other than a Disability as defined in Paragraph 1 hereof), or who is on leave of absence for any purpose, shall not, during the period of any such absence, be deemed, by virtue of such absence alone, to have terminated such Participant's employment, director status or consultancy with the Company or with an Affiliate, except as the Administrator may otherwise expressly provide; provided, however, that, for ISOs, any leave of absence granted by the Administrator of greater than ninety days, unless pursuant to a contract or statute that guarantees the right to reemployment, shall cause such ISO to become a Non-Qualified Option on the 91st day following such leave of absence.

(f) Except as required by law or as set forth in a Participant's Option Agreement, Options granted under the Plan shall not be affected by any change of a Participant's status within or among the Company and any Affiliates, so long as the Participant continues to be an Employee, director or Consultant of the Company or any Affiliate.

14. EFFECT ON OPTIONS OF TERMINATION OF SERVICE FOR CAUSE.

Except as otherwise provided in a Participant's Option Agreement, the following rules apply if the Participant's service (whether as an Employee, director or Consultant) with the Company or an Affiliate is terminated for Cause prior to the time that all his or her outstanding Options have been exercised:

(a) All outstanding and unexercised Options as of the time the Participant is notified his or her service is terminated for Cause will immediately be forfeited.

(b) Cause is not limited to events which have occurred prior to a Participant's termination of service, nor is it necessary that the Administrator's finding of Cause occur prior to termination. If the Administrator determines, subsequent to a Participant's termination of service but prior to the exercise of an Option, that either prior or subsequent to the Participant's

termination the Participant engaged in conduct which would constitute Cause, then the right to exercise any Option is forfeited.

15. EFFECT ON OPTIONS OF TERMINATION OF SERVICE FOR DISABILITY.

(a) Except as otherwise provided in a Participant's Option Agreement, a Participant who ceases to be an Employee, director or Consultant of the Company or of an Affiliate by reason of Disability may exercise any Option granted to such Participant:

(i) To the extent that the Option has become exercisable but has not been exercised on the date of the Participant's termination of service due to Disability; and

(ii) In the event rights to exercise the Option accrue periodically, to the extent of a pro rata portion through the date of the Participant's termination of service due to Disability of any additional vesting rights that would have accrued on the next vesting date had the Participant not become Disabled. The proration shall be based upon the number of days accrued in the current vesting period prior to the date of the Participant's termination of service due to Disability.

(b) A Disabled Participant may exercise the Option only within the period ending one year after the date of the Participant's termination of service due to Disability, notwithstanding that the Participant might have been able to exercise the Option as to some or all of the Shares on a later date if the Participant had not been terminated due to Disability and had continued to be an Employee, director or Consultant or, if earlier, within the originally prescribed term of the Option. For Options granted to California Participants, a Participant may exercise such rights for at least six (6) months from the date of termination of service due to Disability.

(c) The Administrator shall make the determination both of whether Disability has occurred and the date of its occurrence (unless a procedure for such determination is set forth in another agreement between the Company and such Participant, in which case such procedure shall be used for such determination). If requested, the Participant shall be examined by a physician selected or approved by the Administrator, the cost of which examination shall be paid for by the Company.

16. EFFECT ON OPTIONS OF DEATH WHILE AN EMPLOYEE, DIRECTOR OR CONSULTANT.

(a) Except as otherwise provided in a Participant's Option Agreement in the event of the death of a Participant while the Participant is an Employee, director or Consultant of the Company or of an Affiliate, such Option may be exercised by the Participant's Survivors:

(i) To the extent that the Option has become exercisable but has not been exercised on the date of death; and

(ii) In the event rights to exercise the Option accrue periodically, to the extent of a pro rata portion through the date of death of any additional vesting

rights that would have accrued on the next vesting date had the Participant not died. The proration shall be based upon the number of days accrued in the current vesting period prior to the Participant's date of death.

(b) If the Participant's Survivors wish to exercise the Option, they must take all necessary steps to exercise the Option within one year after the date of death of such Participant, notwithstanding that the decedent might have been able to exercise the Option as to some or all of the Shares on a later date if he or she had not died and had continued to be an Employee, director or Consultant or, if earlier, within the originally prescribed term of the Option. For Options granted to California Participants, the Participant's Survivors must be allowed to take all necessary steps to exercise the Option for at least six (6) months from the date of death of such Participant.

17. EFFECT OF TERMINATION OF SERVICE ON SHARE GRANTS AND SHARE-BASED AWARDS.

In the event of a termination of service (whether as an Employee, director or Consultant) with the Company or an Affiliate for any reason before the Participant has accepted a Share Grant or a Share-Based Award and paid the purchase price, if required, such grant shall terminate.

For purposes of this Paragraph 17 and Paragraph 18 below, a Participant to whom a Share Grant or a Share-Based Award has been issued under the Plan who is absent from work with the Company or with an Affiliate because of temporary disability (any disability other than a Disability as defined in Paragraph 1 hereof), or who is on leave of absence for any purpose, shall not, during the period of any such absence, be deemed, by virtue of such absence alone, to have terminated such Participant's employment, director status or consultancy with the Company or with an Affiliate, except as the Administrator may otherwise expressly provide.

In addition, for purposes of this Paragraph 17 and Paragraph 18 below, any change of employment or other service within or among the Company and any Affiliates shall not be treated as a termination of employment, director status or consultancy so long as the Participant continues to be an Employee, director or Consultant of the Company or any Affiliate.

18. EFFECT ON SHARE GRANTS OF TERMINATION OF SERVICE OTHER THAN FOR CAUSE OR DEATH OR DISABILITY.

Except as otherwise provided in a Participant's Share Grant Agreement, in the event of a termination of service (whether as an Employee, director or Consultant), other than termination for Cause, Disability, or death, for which events there are special rules in Paragraphs 19, 20, and 21, respectively, before all forfeiture provisions or Company rights of repurchase shall have lapsed, then the Company shall have the right to cancel or repurchase that number of Shares subject to a Share Grant as to which the Company's forfeiture or repurchase rights have not lapsed.

19. EFFECT ON SHARE GRANTS OF TERMINATION OF SERVICE FOR CAUSE.

Except as otherwise provided in a Participant's Share Grant Agreement, the following rules apply if the Participant's service (whether as an Employee, director or Consultant) with the Company or an Affiliate is terminated for Cause:

(a) All Shares subject to any Share Grant that remain subject to forfeiture provisions or as to which the Company shall have a repurchase right shall be immediately forfeited to the Company as of the time the Participant is notified his or her service is terminated for Cause.

(b) Cause is not limited to events which have occurred prior to a Participant's termination of service, nor is it necessary that the Administrator's finding of Cause occur prior to termination. If the Administrator determines, subsequent to a Participant's termination of service, that either prior or subsequent to the Participant's termination the Participant engaged in conduct which would constitute Cause, then all Shares subject to any Share Grant that remained subject to forfeiture provisions or as to which the Company had a repurchase right on the date of termination shall be immediately forfeited to the Company.

20. EFFECT ON SHARE GRANTS OF TERMINATION OF SERVICE FOR DISABILITY.

Except as otherwise provided in a Participant's Share Grant Agreement, the following rules apply if a Participant ceases to be an Employee, director or Consultant of the Company or of an Affiliate by reason of Disability: to the extent the forfeiture provisions or the Company's rights of repurchase have not lapsed on the date of Disability, they shall be exercisable; provided, however, that in the event such forfeiture provisions or rights of repurchase lapse periodically, such provisions or rights shall lapse to the extent of a pro rata portion of the Shares subject to such Share Grant through the date of Disability as would have lapsed had the Participant not become Disabled. The proration shall be based upon the number of days accrued prior to the date of Disability.

The Administrator shall make the determination both as to whether Disability has occurred and the date of its occurrence (unless a procedure for such determination is set forth in another agreement between the Company and such Participant, in which case such procedure shall be used for such determination). If requested, the Participant shall be examined by a physician selected or approved by the Administrator, the cost of which examination shall be paid for by the Company.

21. EFFECT ON SHARE GRANTS OF DEATH WHILE AN EMPLOYEE, DIRECTOR OR CONSULTANT.

Except as otherwise provided in a Participant's Share Grant Agreement, the following rules apply in the event of the death of a Participant while the Participant is an Employee, director or Consultant of the Company or of an Affiliate: to the extent the forfeiture provisions or the Company's rights of repurchase have not lapsed on the date of death, they shall be exercisable; provided, however, that in the event such forfeiture provisions or rights of repurchase lapse periodically, such provisions or rights shall lapse to the extent of a pro rata

portion of the Shares subject to such Share Grant through the date of death as would have lapsed had the Participant not died. The proration shall be based upon the number of days accrued prior to the Participant's date of death.

22. PURCHASE FOR INVESTMENT.

Unless the offering and sale of the Shares shall have been effectively registered under the Securities Act, the Company shall be under no obligation to issue Shares under the Plan unless and until the following conditions have been fulfilled:

(a) The person who receives a Share Right shall warrant to the Company, prior to the receipt of Shares, that such person is acquiring such Shares for his or her own account, for investment, and not with a view to, or for sale in connection with, the distribution of any such Shares, in which event the person acquiring such Shares shall be bound by the provisions of the following legend (or a legend in substantially similar form) (which shall be endorsed upon any certificate evidencing the Shares that the Company may issue pursuant to such exercise or such grant):

> "The shares represented by this certificate have been taken for investment and they may not be sold or otherwise transferred by any person, including a pledgee, unless (1) either (a) a Registration Statement with respect to such shares shall be effective under the Securities Act of 1933, as amended, or (b) the Company shall have received an opinion of counsel satisfactory to it that an exemption from registration under such Act is then available, and (2) there shall have been compliance with all applicable state securities laws."

(b) At the discretion of the Administrator, the Company shall have received an opinion of its counsel that the Shares may be issued in compliance with the Securities Act without registration thereunder.

23. DISSOLUTION OR LIQUIDATION OF THE COMPANY.

Upon the dissolution or liquidation of the Company, all Options granted under this Plan which as of such date shall not have been exercised and all Share Grants and Share-Based Awards which have not been accepted, to the extent required under the applicable Agreement, will terminate and become null and void; provided, however, that if the rights of a Participant or a Participant's Survivors have not otherwise terminated and expired, the Participant or the Participant's Survivors will have the right immediately prior to such dissolution or liquidation to exercise or accept any Share Right to the extent that the Share Right is exercisable or subject to acceptance as of the date immediately prior to such dissolution or liquidation. Upon the dissolution or liquidation of the Company, any outstanding Share-Based Awards shall immediately terminate unless otherwise determined by the Administrator or specifically provided in the applicable Agreement.

24. ADJUSTMENTS.

Upon the occurrence of any of the following events, a Participant's rights with respect to any Share Right granted to him or her hereunder shall be adjusted as hereinafter provided, unless otherwise specifically provided in a Participant's Agreement:

(a) Share Dividends and Share Splits. If (i) the Common Shares shall be subdivided or combined into a greater or smaller number of shares or if the Company shall issue any Common Shares as a share dividend on its outstanding Common Shares, or (ii) additional shares or new or different shares or other securities of the Company or other non-cash assets are distributed with respect to such Common Shares, each Share Right and the number of Common Shares deliverable thereunder shall be appropriately increased or decreased proportionately, and appropriate adjustments shall be made including, in the exercise or purchase price per share, to reflect such events. The number of Shares subject to the limitations in Paragraph 3(a) shall also be proportionately adjusted upon the occurrence of such events.

(b) Corporate Transactions. If the Company is to be consolidated with or acquired by another entity in a merger, amalgamation, consolidation, or sale of all or substantially all of the Company's assets other than a transaction to merely change the state of incorporation (a "Corporate Transaction"), the Administrator or the board of directors of any entity assuming the obligations of the Company hereunder (the "Successor Board"), shall, as to outstanding Options, either (i) make appropriate provision for the continuation of such Options by substituting on an equitable basis for the Shares then subject to such Options either the consideration payable with respect to the outstanding Common Shares in connection with the Corporate Transaction or securities of any successor or acquiring entity; or (ii) upon written notice to the Participants, provide that such Options must be exercised (either (A) to the extent then exercisable or, (B) at the discretion of the Administrator, any such Options being made partially or fully exercisable for purposes of this Subparagraph), within a specified number of days of the date of such notice, at the end of which period such Options which have not been exercised shall terminate; or (iii) terminate such Options in exchange for payment of an amount equal to the consideration payable upon consummation of such Corporate Transaction to a holder of the number of Common Shares into which such Option would have been exercisable (either (A) to the extent then exercisable or, (B) at the discretion of the Administrator, any such Options being made partially or fully exercisable for purposes of this Subparagraph) less the aggregate exercise price thereof. For purposes of determining the payments to be made pursuant to Subclause (iii) above, in the case of a Corporate Transaction the consideration for which, in whole or in part, is other than cash, the consideration other than cash shall be valued at the fair value thereof as determined in good faith by the Board of Directors.

With respect to outstanding Share Grants, the Administrator or the Successor Board shall 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 such Share Grants either the consideration payable with respect to such outstanding Shares in connection with the Corporate Transaction or securities of any successor or acquiring entity. In lieu of the foregoing, in connection with any Corporate Transaction, the Administrator may provide that, upon consummation of the Corporate Transaction, each outstanding Share Grant shall be terminated in exchange for payment of an amount equal to the consideration payable upon consummation of

such Corporate Transaction to a holder of the number of Common Shares comprising such Share Grant (to the extent such Share Grant is no longer subject to any forfeiture or repurchase rights then in effect or, at the discretion of the Administrator, all forfeiture and repurchase rights being waived upon such Corporate Transaction).

In taking any of the actions permitted under this Paragraph 24(b), the Administrator shall not be obligated by the Plan to treat all Share Rights, all Share Rights held by a Participant, or all Share Rights of the same type, identically.

(c) Recapitalization or Reorganization. In the event of a recapitalization or reorganization of the Company other than a Corporate Transaction pursuant to which securities of the Company or of another corporation are issued with respect to the outstanding Common Shares, a Participant upon exercising an Option or accepting a Share Grant after the recapitalization or reorganization shall be entitled to receive for the price paid upon such exercise or acceptance if any, the number of replacement securities which would have been received if such Option had been exercised or Share Grant accepted prior to such recapitalization or reorganization.

(d) Adjustments to Share-Based Awards. Upon the happening of any of the events described in Subparagraphs (a), (b) or (c) above, any outstanding Share-Based Award shall be appropriately adjusted to reflect the events described in such Subparagraphs. The Administrator or the Successor Board shall determine the specific adjustments to be made under this Paragraph 24, including, but not limited to the effect of any Corporate Transaction and, subject to Paragraph 4, its determination shall be conclusive.

(e) Modification of Options. Notwithstanding the foregoing, any adjustments made pursuant to Subparagraph (a), (b) or (c) above with respect to Options shall be made only after the Administrator determines whether such adjustments would (i) constitute a "modification" of any ISOs (as that term is defined in Section 424(h) of the Code) or (ii) cause any adverse tax consequences for the holders of Options, including, but not limited to, pursuant to Section 409A of the Code. If the Administrator determines that such adjustments made with respect to Options would constitute a modification or other adverse tax consequence, it may refrain from making such adjustments, unless the holder of an Option specifically agrees in writing that such adjustment be made and such writing indicates that the holder has full knowledge of the consequences of such "modification" on his or her income tax treatment with respect to the Option. This paragraph shall not apply to the acceleration of the vesting of any ISO that would cause any portion of the ISO to violate the annual vesting limitation contained in Section 422(d) of the Code, as described in Paragraph 6(b)(iv).

25. ISSUANCES OF SECURITIES.

Except as expressly provided herein, no issuance by the Company of shares of any class, or securities convertible into shares of any class, shall affect, and no adjustment by reason thereof shall be made with respect to, the number or price of shares subject to Share Rights. Except as expressly provided herein, no adjustments shall be made for dividends paid in cash or in property (including without limitation, securities) of the Company prior to any issuance of Shares pursuant to a Share Right.

26. FRACTIONAL SHARES.

No fractional shares shall be issued under the Plan and the person exercising a Share Right shall receive from the Company cash in lieu of such fractional shares equal to the Fair Market Value thereof.

27. CONVERSION OF ISOs INTO NON-QUALIFIED OPTIONS; TERMINATION OF ISOs.

The Administrator, at the written request of any Participant, may in its discretion take such actions as may be necessary to convert such Participant's ISOs (or any portions thereof) that have not been exercised on the date of conversion into Non-Qualified Options at any time prior to the expiration of such ISOs, regardless of whether the Participant is an Employee of the Company or an Affiliate at the time of such conversion. At the time of such conversion, the Administrator (with the consent of the Participant) may impose such conditions on the exercise of the resulting Non-Qualified Options as the Administrator in its discretion may determine, provided that such conditions shall not be inconsistent with this Plan. Nothing in the Plan shall be deemed to give any Participant the right to have such Participant's ISOs converted into Non-Qualified Options, and no such conversion shall occur until and unless the Administrator takes appropriate action. The Administrator, with the consent of the Participant, may also terminate any portion of any ISO that has not been exercised at the time of such conversion.

28. WITHHOLDING.

In the event that any federal, state, or local income taxes, employment taxes, Federal Insurance Contributions Act ("F.I.C.A.") withholdings or other amounts are required by applicable law or governmental regulation to be withheld from the Participant's salary, wages or other remuneration in connection with the issuance of a Share Right or Shares under the Plan or for any other reason required by law, the Company may withhold from the Participant's compensation, if any, or may require that the Participant advance in cash to the Company, or to any Affiliate of the Company which employs or employed the Participant, the statutory minimum amount of such withholdings unless a different withholding arrangement, including the use of shares of the Company's Common Shares or a promissory note, is authorized by the Administrator (and permitted by law). For purposes hereof, the fair market value of the shares withheld for purposes of payroll withholding shall be determined in the manner set forth under the definition of Fair Market Value provided in Paragraph 1 above, as of the most recent practicable date prior to the date of exercise. If the Fair Market Value of the shares withheld is less than the amount of payroll withholdings required, the Participant may be required to advance the difference in cash to the Company or the Affiliate employer. The Administrator in its discretion may condition the exercise of an Option for less than the then Fair Market Value on the Participant's payment of such additional withholding.

29. NOTICE TO COMPANY OF DISQUALIFYING DISPOSITION.

Each Employee who receives an ISO must agree to notify the Company in writing immediately after the Employee makes a Disqualifying Disposition of any Shares acquired pursuant to the exercise of an ISO. A Disqualifying Disposition is defined in Section 424(c) of

the Code and includes any disposition (including any sale or gift) of such Shares before the later of (a) two years after the date the Employee was granted the ISO, or (b) one year after the date the Employee acquired Shares by exercising the ISO, except as otherwise provided in Section 424(c) of the Code. If the Employee has died before such Shares are sold, these holding period requirements do not apply and no Disqualifying Disposition can occur thereafter.

30. TERMINATION OF THE PLAN.

The Plan will terminate on December 16, 2025, the date which is ten years from the earlier of the date of its adoption by the Board of Directors and the date of its approval by the shareholders of the Company. The Plan may be terminated at an earlier date by vote of the shareholders or the Board of Directors of the Company; provided, however, that any such earlier termination shall not affect any Agreements executed prior to the effective date of such termination. Termination of the Plan shall not affect any Share Rights theretofore granted.

31. AMENDMENT OF THE PLAN AND AGREEMENTS.

The Plan may be amended by the shareholders of the Company. The Plan may also be amended by the Administrator, including, without limitation, to the extent necessary to qualify any or all outstanding Share Rights granted under the Plan or Share Rights to be granted under the Plan for favorable federal income tax treatment as may be afforded incentive stock options under Section 422 of the Code (including deferral of taxation upon exercise), and to the extent necessary to qualify the Shares issuable under the Plan for listing on any national securities exchange or quotation in any national automated quotation system of securities dealers. Any amendment approved by the Administrator which the Administrator determines is of a scope that requires shareholder approval shall be subject to obtaining such shareholder approval. Any modification or amendment of the Plan shall not, without the consent of a Participant, adversely affect his or her rights under a Share Right previously granted to him or her. With the consent of the Participant affected, the Administrator may amend outstanding Agreements in a manner which may be adverse to the Participant but which is not inconsistent with the Plan. In the discretion of the Administrator, outstanding Agreements may be amended by the Administrator in a manner which is not adverse to the Participant.

32. EMPLOYMENT OR OTHER RELATIONSHIP.

Nothing in this Plan or any Agreement shall be deemed to prevent the Company or an Affiliate from terminating the employment, consultancy or director status of a Participant, nor to prevent a Participant from terminating his or her own employment, consultancy or director status or to give any Participant a right to be retained in employment or other service by the Company or any Affiliate for any period of time.

33. GOVERNING LAW.

This Plan shall be construed and enforced in accordance with the law of Bermuda.

KINIKSA PHARMACEUTICALS, LTD.

Share Option Grant Notice
Share Option Grant under the Company's
2015 Equity Incentive Plan

1. Name and Address of Participant:

2. Date of Option Grant:

3. Type of Grant:

4. Maximum Number of Shares for which this Option is exercisable:

5. Exercise (purchase) price per share:

6. Option Expiration Date:

7. Vesting Start Date:

8. Vesting Schedule: This Option shall become exercisable (and the Shares issued upon exercise shall be vested) as follows provided the Participant is an Employee, director or Consultant of the Company or of an Affiliate on the applicable vesting date:

On the first anniversary of the Vesting Start Date	25% of the Shares
Thereafter, on each one month anniversary of the Vesting Start Date	an additional 2.0833% of the Shares

In accordance with the above, all the Shares subject to the Option shall be fully vested 4 years from the Vesting Start Date.

The foregoing rights are cumulative and are subject to the other terms and conditions of this Share Option Grant Notice, and the terms of the Share Option Agreement attached hereto, and the Company's 2015 Equity Incentive Plan.

The Company and the Participant acknowledge receipt of this Share Option Grant Notice and agree to the terms of the Share Option Agreement attached hereto and incorporated by reference herein, the Company's 2015 Equity Incentive Plan and the terms of this Share Option Grant Notice as set forth above.

KINIKSA PHARMACEUTICALS, LTD.

By: _____
Name: _____
Title: _____

_____
Participant

Exhibit 10.10

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

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.

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Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

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Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

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.

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

"***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

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(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

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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

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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

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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

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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) [***] 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

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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,

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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

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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).

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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

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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 [***]

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

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

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)
[***]	$ [***]
[***]	$ [***]

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

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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.

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

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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

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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

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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

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.

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

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(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

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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

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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

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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

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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

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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

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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

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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

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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

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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 [***], 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) [***], 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,

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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

Schedule A

Purchased Assets

1. **Acquired Patent Rights**

[***]

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.

2. Acquired Know-How

[***]

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.

3. **Assigned Contracts**

[***]

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.

4. Inventory

A. [***]

[REDACTED]

B. [***]

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.

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

[***]

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.

Schedule G

Completion of Technology Transfer

[***]

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.

Schedule H

Individuals for Purposes of Knowledge

[***]

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.

Schedule I

Exceptions to Biogen Obligations/Representations in Sections 2.6.6 and 6.2.4(g) and (l)

[***]

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.

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:

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

ATTACHMENT A

Assigned Patents

[***]

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.

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:

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.

"***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 F-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.

Exhibit 10.11

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

LICENSE AGREEMENT

By and Between

REGENERON PHARMACEUTICALS, INC.

and

KINIKSA PHARMACEUTICALS, LTD.

Dated as of September 25, 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.

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SCHEDULES

LICENSE AGREEMENT

THIS LICENSE AGREEMENT ("Agreement"), dated as of September 25, 2017 (the "Effective Date"), is by and between REGENERON PHARMACEUTICALS, INC., a corporation organized under the laws of New York and having an address at 777 Old Saw Mill River Road, Tarrytown, New York 10591 ("Regeneron"), and KINIKSA PHARMACEUTICALS, LTD., a Bermuda exempted company and having an address at Clarendon House, 2 Church Street, Hamilton HM 11, Bermuda ("Kiniksa") (with each of Regeneron and Kiniksa referred to herein individually as a "Party" and collectively as the "Parties").

WHEREAS, Kiniksa possesses knowledge and expertise in, and resources for, developing and commercializing pharmaceutical products in the Territory;

WHEREAS, Regeneron wishes to grant Kiniksa the exclusive right to develop and commercialize the Product in the Kiniksa Field in the Territory on the terms set forth herein; and

WHEREAS, Regeneron desires to supply Kiniksa with the Product for development and commercialization in the Kiniksa Field in the Territory on the terms set forth herein.

NOW, THEREFORE, in consideration of the following mutual promises and obligations, and for other good and valuable consideration, the adequacy and sufficiency of which are hereby acknowledged, the Parties agree as follows:

ARTICLE 1
DEFINITIONS

Capitalized terms used in this Agreement, whether used in the singular or plural, except as expressly set forth herein, shall have the meanings set forth below:

1.1 "Affiliate" shall mean, with respect to any Person, another Person that controls, is controlled by, or is under common control with such Person. A Person shall control another Person if such Person possesses, directly or indirectly, the power to direct or cause the direction of the management and policies of such other Person, whether through the ownership of voting securities, by contract, or otherwise. Without limiting the generality of the

foregoing, a Person shall control another Person if any of the following conditions are met: (a) in the case of corporate entities, direct or indirect ownership of more than fifty percent (50%) of the stock or shares having the right to vote for the election of directors, and (b) in the case of non-corporate entities, direct or indirect ownership of more than fifty percent (50%) of the equity interest with the power to direct the management and policies of such non-corporate entities. The Parties acknowledge that in the case of certain entities organized under the laws of certain countries outside of the United States, the maximum percentage ownership permitted by law for a foreign investor may be less than fifty percent (50%), and that in such case such lower percentage

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shall be substituted in the preceding sentence; *provided that* such foreign investor has the power to direct the management and policies of such entity. Notwithstanding the foregoing, neither [***] shall be considered an Affiliate of Kiniksa.

1.2 "Agreement" shall have the meaning set forth in the introductory paragraph, and includes all Schedules and Exhibits attached hereto.

1.3 "Alliance Manager" has the meaning set forth in Section 4.3 (Alliance Management).

1.4 "[***]" shall mean [***].

1.5 "[***]" shall mean [***].

1.6 "Anticipated First Commercial Sale" shall mean, with respect to a Product in the Kiniksa Field in a particular country in the Territory, the expected date of First Commercial Sale of such Product in the Kiniksa Field in such country.

1.7 "Anticipated Supply Shortage" has the meaning set forth in Section 8.13 (Notification and Discussion of Supply Issues).

1.8 "Anti-Corruption Laws" shall mean all Applicable Laws regarding public or private-sector corruption, bribery, kickbacks, speed or facilitation payments, ethical business conduct, money laundering, embezzlement, political contributions, gifts, gratuities, expenses, entertainment, hospitalities, agency relationships, commissions, lobbying, books and records, and financial controls, including the FCPA, the U.S. Travel Act, the UK Bribery Act 2010, and other anti-corruption laws, in each case, as amended.

1.9 "Applicable Law" shall mean any federal, state, local, national, and supra-national law, rule, regulation, statute, treaty, or ordinance of any Governmental Authority, including any rules, regulations, guidelines, or other requirements of any Regulatory Authority, in each case, that may be in effect from time to time and applicable to a particular activity under this Agreement.

1.10 "Approval" shall mean, with respect to a pharmaceutical product, any approval (including any Marketing Approval or Pricing Approval), registration, license, or authorization from any Regulatory Authority or other Governmental Authority that is required for the Development, Manufacture, or Commercialization of such product in a country or other regulatory jurisdiction anywhere in the Territory, and shall include any approval, registration, license, or authorization granted in connection with any Registration Filing.

1.11 "Audited Party" has the meaning set forth in Section 11.1 (Books and Records).

1.12 "Auditing Party" has the meaning set forth in Section 11.1 (Books and Records).

1.13 "Big Four" has the meaning set forth in Section 11.2.1 (Right to Audit).

1.14 "BLA" shall mean a biologics license application, supplemental biologics application, or biologics application amendment, in each case, as described in the FDA regulations, including all amendments and supplements to the application, and any equivalent filing with any Regulatory Authority in a country or regulatory jurisdiction to obtain Marketing Approval to Commercialize a pharmaceutical product in such country or jurisdiction.

1.15 "BLA Filing" shall mean the complete submission and acceptance by the applicable Regulatory Authority of a BLA for filing; *provided that* such Regulatory Authority has not issued a refusal to file letter or a letter identifying deficiencies for which the Regulatory Authority will suspend its review following submission of such filing.

1.16 "Business Day" shall mean any day other than a Saturday, a Sunday, or other day on which commercial banks in New York, New York, or Boston, Massachusetts are authorized or required by law to remain closed.

1.17 "CAPS" shall mean Cryopyrin-Associated Autoinflammatory Syndrome.

1.18 "CDA" has the meaning set forth in Section 13.1 (Confidential Information).

1.19 "Certain Shared Commercial Expenses [***]" has the meaning set forth in Section 9.5.1(d) ([***] Inclusion).

1.20 "Challenge" has the meaning set forth in Section 16.7 (Termination for IP Challenge).

1.21 "Challenged Patent Right" has the meaning set forth in Section 16.7 (Termination for IP Challenge).

1.22 "Change of Control" shall mean, with respect to a Party, (a) a merger or consolidation of such Party with a Third Party that results in the voting securities of such Party outstanding immediately prior thereto, or any securities into which such voting securities have been converted or exchanged, ceasing to represent more than fifty percent (50%) of the combined voting power of the surviving entity or the parent of the surviving entity immediately after such merger or consolidation, or (b) a transaction or series of related transactions in which a Third Party, together with its Affiliates, becomes the beneficial owner of more than fifty percent (50%) of the combined voting power of the outstanding securities of such Party, or (c) the sale or other transfer to a Third Party of all or substantially all of such Party's assets or business; *provided*, *however*, that a "Change of Control" will not include (i) any public offering of securities or any transaction or series of transactions in which a financial investor becomes the beneficial owner of fifty percent (50%) or more of the combined voting power, or (ii) any transaction principally undertaken for *bona fide* equity financing purposes with financial investors (excluding a financial investor that is the investment entity of a pharmaceutical company) in which (A) cash is received by Kiniksa or any successor entity or indebtedness of Kiniksa is cancelled or converted, and (B) no direct, indirect, or conditional license or transfer of any rights to Intellectual Property Rights of Kiniksa occurs or is in any way contemplated in connection therewith.

1.23 "Clinical Supply Requirements" shall mean, with respect to the Product and placebo, the quantities of such Product and placebo ordered by Kiniksa for Development purposes under the Clinical Supply Agreement.

1.24 "Commercial Clinical Studies" shall mean clinical studies solely intended to collect information to support pricing activities (*e.g.*, outcomes data to support evidence based pricing negotiations with payors) for any Product in the Kiniksa Field in the Territory. For clarity, Commercial Clinical Studies shall exclude any study that is intended to support the Approval of additional labeled Indications, line extensions, or formulations of any Product.

1.25 "Commercial Information" shall mean, with respect to a Product, information relating to the pricing, reimbursement, marketing, promotion, distribution, offering for sale, or selling of such Product (including information pertaining to, or submitted in support of, all Pricing Approvals), including information about pharmaco-economic studies justifying pricing, analysis of market sales and prescription data, analysis of competitive products and environment, product positioning (including unique selling proposition and understanding of competitors' positioning strategies) and promotional strategies (including promotional materials); virtual product and clinical support information (web page); ongoing medical education strategies; and strategies used for building relationships with health insurance and managed care entities.

1.26 "Commercial Overhead Charge" shall mean, on a country-by-country and year-by-year basis, beginning on the later of (a) the date of [***], or (b) [***], an amount to cover Kiniksa's FTE Costs for Product management, contract administration, fleet administration, sales operations (*e.g.*, physician targeting, commission administration), sales material fulfillment, advertising operations, convention planning, seeking Pricing Approvals, maintenance of Approvals, surveys (but only to the extent not required by a Regulatory Authority to obtain any Approval), registries (but only to the extent not required by a Regulatory Authority to obtain any Approval) and Commercial Clinical Studies, scientific publications, health outcomes, pricing and reimbursement activities, procurement services, customer service, patient assistance programs, and trade administration, in each case, to the extent attributable to the Commercialization of a Product in the Kiniksa Field in such country and not included in Field Force Costs, Other Shared Expenses, or otherwise in Shared Commercial Expenses. For the avoidance of doubt, the Commercial Overhead Charge shall not include the costs of personnel for personnel functions not directly involved in the Commercialization of a Product.

1.27 "Commercial Plan" shall mean, with respect to a Product, the [***] rolling plan and budget that describes the significant Commercialization activities (including significant pre-launch and launch activities) planned to be undertaken by Kiniksa for such Product in the Kiniksa Field in the Territory and the associated budget for such Commercialization activities, including a summary of the following with respect to the Product: (a) anticipated major advertising, public relations, and patient advocacy or education programs, (b) strategies for Promotion, (c) pricing strategy, (d) market access strategy, (e)

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reimbursement and patient support programs, (f) competitive positioning, (g) Anticipated First Commercial Sale, and (h) market and sales forecasts.

1.28 "Commercial Quality Agreement" has the meaning set forth in Section 8.6 (Quality Agreements).

1.29 "Commercial Supply Agreement" has the meaning set forth in Section 8.5 (Commercial Supply Agreement).

1.30 "Commercial Supply Requirements" shall mean, with respect to a Product, the quantities of such Product ordered by Kiniksa for Commercialization in the Kiniksa Field in the Territory under the Commercial Supply Agreement, including, as applicable, quantities required for pre-launch stockpiling, samples, Commercial Clinical Studies, safety stock, and provision of Product for free commercial use.

1.31 "Commercialize" or "Commercialization" shall mean, with respect to a product, any and all activities directed to marketing, distributing, market access, detailing, promoting, pricing, reimbursing, offering for sale, selling, or importing, such product in the Territory, including market research, obtaining Pricing Approvals, customer service, marketing and educational activities, surveys, registries and Commercial Clinical Studies, and interacting with Regulatory Authorities and other Governmental Authorities following Marketing Approval in the applicable country or regulatory jurisdiction pertaining to Indications for which Marketing Approval has been obtained and post-Marketing Approval pharmacovigilance (for clarity, excluding pharmacovigilance for clinical trials under the Development Plan). Commercialize and Commercialization activities shall not include Manufacture or Manufacturing activities or Development or Development activities.

1.32 "Commercially Reasonable Efforts" shall mean, with respect to the efforts to be expended by a Party with respect to any objective, commercially reasonable, diligent, good faith efforts to accomplish such objective under this Agreement, which efforts shall be consistent with the exercise of prudent scientific and business judgment in accordance with the efforts that [***] would normally use to accomplish a similar objective under similar circumstances. It being understood and agreed that such efforts shall be [***]. Commercially Reasonable Efforts will be determined on a market-by-market basis in view of conditions prevailing at the time, and evaluated taking into account all relevant factors, including the efficacy, safety, product profile, profitability, sales potential, return on investment, intellectual property considerations, anticipated and actual regulatory authority approved labeling, anticipated and actual pricing and reimbursement, competitiveness of such Product, or alternative products that are in the marketplace or under development by Third Parties and other technical, scientific, legal, medical marketing, and competitiveness factors. In determining whether a Party has used Commercially Reasonable Efforts, [***].

1.33 "Confidential Information" has the meaning set forth in Section 13.1 (Confidential Information).

1.34 "Contract Year" shall mean the period beginning on the Effective Date and ending on December 31, 2017, and each succeeding twelve (12) month period thereafter during the Term (except that the last Contract Year shall end on the effective date of any termination or expiration of this Agreement).

1.35 "Control" shall mean, with respect to any material, Confidential Information, Intellectual Property Right, or Trademark that a Party (a) owns such material, Confidential Information, Intellectual Property Right, or Trademark, or (b) has a license or right to use to such material, Confidential Information, Intellectual Property Right, or Trademark, in each case of (a) or (b), with the ability to grant to the other Party access, a right to use, or a license, or a sublicense (as applicable) to such material, Confidential Information, Intellectual Property Right, or Trademark on the terms and conditions set forth herein, without (i) violating the terms of any agreement with or obligation to any Third Party in existence as of the time such Party or its Affiliates would first be required hereunder to grant the other Party such access, right to use, or (sub)license or (ii) incurring any additional payment obligations to any Third Party; *provided that* any material, Confidential Information, Intellectual Property Right, or Trademark in-licensed by a Party from a Third Party that would otherwise be a Third Party License hereunder, but that the JSC does not approve the entry into pursuant to Section 4.1.3(t) will not be Controlled for purposes of this Agreement. Notwithstanding the foregoing, no Patent Rights, Know-How, or other Intellectual Property Right will be Controlled by either Party hereunder if such Patent Right, Know-How, or other Intellectual Property Right is owned or in-licensed by a Third Party that becomes an Affiliate of such Party after the Effective Date as a result of such Party being acquired by such Third Party, whether by merger, stock purchase, or purchase of assets; *provided that* prior to the date of such transaction, neither such Party nor any of its Affiliates had any rights to any such Patent Right, Know-How, or other Intellectual Property Right. Notwithstanding the foregoing, any such Patent Right, Know-How, or other Intellectual Property Right that is owned or in-licensed by such acquiring Third Party and that is actually used following the date of such transaction by such Third Party or acquired Party in connection with the Development, Manufacture, or Commercialization of any Product or that is invented or created by the acquiring Third Party with the use or incorporation of Patent Rights, Know-How, or other Intellectual Property Rights licensed hereunder will be Controlled by a Party (as an Affiliate of such Third Party) for purposes of this Agreement.

1.36 "Cost of Finishing" shall mean the costs of converting Formulated Bulk Product to Product in finished form (including all packaging and labeling), including vial filling, lyophilizing, device manufacture and assembly, packaging, labeling, and testing Filled Product, and associated freight, insurance, and quality control. If such finishing activities are performed by a Party or its Affiliates, then the Cost of Finishing shall be such Party's actual cost without any profit for such Party or mark-up by such Party. If such finishing activities are performed by a Third Party, then the Cost of Finishing shall be Out-of-Pocket Costs paid by the contracting Party to such Third Party without any mark-up, and will include any fees paid to such Third Party to [***] to the extent [***] the Manufacturing Cost.

1.37 "Cost of Goods Sold" or "COGS" shall mean, for a Quarter, the [***] for Products sold, otherwise distributed for patient use, or that expired in the Territory during such Quarter (and not in the Quarter in which such Product was Manufactured). COGS excludes all costs and expenses included in Other Shared Expenses and Shared Commercial Expenses.

1.38 "Cover," "Covers," or "Covered" means, with respect to a particular subject matter at issue and a claim of a relevant Patent Right, that the unlicensed or unauthorized manufacture, use, sale, offer for sale, or importation of the subject matter would infringe such claim (if in a Patent, or, in the case of claim in a Patent Application, would infringe such claim if it were to issue in a Patent without modification).

1.39 "CPI" shall mean, with respect to personnel located in the U.S., the Consumer Price Index — All Urban Consumers published by the United States Department of Labor, Bureau of Statistics (or its successor equivalent index), and with respect to personnel located outside the U.S., (a) an equivalent index in a foreign country applicable to FTEs in such country, accounting if possible for the area in such country where the personnel are located, or (b) other inflation measure or rate agreed to by the Parties.

1.40 "CPI Adjustment" shall mean the percentage increase or decrease, if any, in the CPI applicable to such personnel for the twelve (12) months ending September 30 of the Contract Year prior to the Contract Year for which the adjustment is being made.

1.41 "CREATE Act" has the meaning set forth in Section 10.7.6 (CREATE Act).

1.42 "Damages" has the meaning set forth in Section 14.1 (Indemnity by Kiniksa).

1.43 "Debarred" has the meaning set forth in Section 12.4.11 (Debarment).

1.44 "Detail" shall mean, with respect to the Product, a selling presentation for such Product by a representative of a Party's sales force, or another employee of such Party who may be deemed to be part of the promotional activities for such Product (*e.g.*, key account manager), in each case, with a healthcare provider who has prescribing authority or is able to influence or is responsible for patient identification or management and is within the designated target audience during which approved uses, safety, effectiveness, contraindications, side effects, warnings, or other relevant characteristics of such Product are discussed in an effort to increase prescribing preferences of such Product for its approved uses.

1.45 "Develop" or "Development" shall mean, with respect to a pharmaceutical product, the following activities undertaken or performed for such product: (a) activities relating to research and pre-clinical and clinical development of such Product, including test method development and stability testing, assay development, toxicology, pharmacology, formulation, statistical analysis, pharmacokinetic studies, data collection and management, clinical studies (including the design of clinical studies and any extensions to clinical studies initiated prior to Marketing Approval or required to maintain

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Marketing Approval) but excluding Commercial Clinical Studies, regulatory affairs, project management, drug safety surveillance activities related to clinical studies, preparation submission and maintenance of Registration Filings, maintenance of Approvals, activities necessary to obtain any Pricing Approval, reimbursement, or listing on health care providers' and payers' formularies, and (b) any other research and development activities with respect to such product, including activities to support the discovery of biomarkers and activities to support new product formulations, delivery technologies, or new Indications, either before or after the First Commercial Sale of such product. Develop and Development activities shall not include Manufacture or Manufacturing activities or Commercialization or Commercialization activities.

1.46 "Development Milestone Event" has the meaning set forth in Section 9.2.1 (Milestone Payments).

1.47 "Development Milestone Payment" has the meaning set forth in Section 9.2.1 (Milestone Payments).

1.48 "Development Plan" shall mean, with respect to a Product, the [***] year rolling plan and budget for the Development of such Product for Commercialization in the Kiniksa Field in the Territory, which plan shall include the following:

(a) the overall strategies and estimated timelines for Developing and submitting Approvals for such Product in the Kiniksa Field in the Territory;

(b) proposed clinical study design, clinical methodology, and monitoring requirements for clinical trials of such Product; and

(c) planned pre-clinical research directed to such Product.

1.49 "Development Quality Agreement" has the meaning set forth in Section 8.6 (Quality Agreements).

1.50 "Development Supply Agreement" has the meaning set forth in Section 8.3 (Development Supply Agreement).

1.51 "DIRA" shall mean Deficiency of the Interleukin-1 Receptor Antagonist.

1.52 "Disclosing Party" has the meaning set forth in Section 13.1 (Confidential Information).

1.53 "EMA" shall mean the European Medicines Agency or any successor agency thereto.

1.54 "Executive Officers" shall mean the Chief Executive Officer of Regeneron and the Chief Executive Officer of Kiniksa, or their respective designees with equivalent decision-making authority with respect to matters under this Agreement.

1.55 "Existing Contracts" has the meaning set forth in Section 3.5.2 (Existing Contracts).

1.56 "Existing Licenses" shall mean [***].

1.57 "Existing Product Formulation" shall mean (a) the 160mg dosage form as Commercialized by Regeneron as of the Effective Date, and (b) the 80mg dosage form, which 80 mg dosage form was previously developed by Regeneron but is not Commercialized by Regeneron as of the Effective Date.

1.58 "Existing Product Trademarks" shall mean those Trademarks Controlled by Regeneron and used in connection with the Commercialization of any Product (a) as of the Effective Date, or (b) in the Regeneron Field prior to the date of receipt of U.S. Marketing Approval. The Existing Product Trademarks as of the Effective Date are set forth on Schedule 1.58.

1.59 "Existing Regeneron Patent Rights" has the meaning set forth in Section 10.7.2 (Existing Regeneron Patent Rights; Costs).

1.60 "FCPA" shall mean the U.S. Foreign Corrupt Practices Act of 1977 (15 U.S.C.§§78dd-1, *et seq.*) as amended.

1.61 "FDA" shall mean the United States Food and Drug Administration and any successor agency thereto.

1.62 "FD&C Act" means the United States Federal 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).

1.63 "Field Force Cost" shall mean, in any country or Region in the Territory, the product of (a) the number of Kiniksa FTEs conducting Details, performing account management, medical science liaison, or medical affairs functions with respect to a Product, and (b) the applicable Field Force FTE Rate. For the avoidance of doubt, the activities of Third Party contract personnel shall be charged as Out-of-Pocket Costs under Section 1.189(b) (Shared Commercial Expenses) and not included in Field Force Costs.

1.64 "Field Force FTE Rates" shall mean, on a country-by-country or Region-by-Region basis (determined based on the location of the field force representative), a rate or rates based upon the [***]. The Field Force FTE Rate shall be inclusive of Out-of-Pocket Costs and other expenses for the employee providing the services, including [***] to the extent attributable to the field force.

1.65 "Filled Product" shall mean a Product (a) in the Existing Product Formulation in its filled form in unlabeled vials, and (b) in any other form mutually agreed to be supplied by Regeneron under the Supply Agreements in filled form, in each case, ready to be finished.

1.66 "Fill/Finish Technology Transfer" has the meaning set forth in Section 8.14.2 (Fill/Finish Transfer).

1.67 "Financial Dispute" shall mean any dispute related to (a) a Party's method of calculation of any element included in the Profit Split Arrangement, including any apportionment of costs and expenses included therein, whether before or after the date of receipt of U.S. Marketing Approval, or (b) a Party's method of calculation of the Third Party Proceeds.

1.68 "Financial Expert" has the meaning set forth in Section 17.1.2 (Financial Disputes).

1.69 "First Commercial Sale" shall mean, with respect to a Product in a country, the first commercial sale of such Product for end use or consumption to a Third Party in such country following receipt of Marketing Approval and Pricing Approval (where applicable) in such country. Sales or other distribution for test marketing or clinical trial purposes or compassionate or similar use, including extended access programs, shall not constitute a First Commercial Sale.

1.70 "Force Majeure" has the meaning set forth in ARTICLE 15 (Force Majeure).

1.71 "Formulated Bulk Product" shall mean (a) the Existing Product Formulation formulated into solution or in a lyophilized form, and (b) any other form of any Product to be supplied by Regeneron under a Supply Agreement that is ready for storage or shipment to a manufacturing facility to allow processing into the final dosage form.

1.72 "Formulated Bulk Technology Transfer" has the meaning set forth in Section 8.15.1 (Transfers).

1.73 "Formulation Development Activities" has the meaning set forth in Section 4.1.3(x) (Duties of the JSC).

1.74 "FTE" shall mean a full time equivalent employee (*i.e.*, one fully-committed or multiple partially-committed employees aggregating to one full-time employee) employed by a Party (or its Affiliate) who performs work related to the Development, Manufacture, or Commercialization of any Product, or other activities with respect to any Product under this Agreement, in each case, for which a Party is entitled to reimbursement under this Agreement or that may be billed under the Profit Split Arrangement, with such commitment of time and effort to constitute one employee performing such work on a full-time basis, which for purposes hereof shall be [***] hours per year.

1.75 "FTE Cost" shall mean the product of (a) the number of FTEs performing activities for which a Party entitled to reimbursement to under this Agreement or that may be billed under the Profit Split Arrangement, and (b) the applicable FTE Rate for such FTE.

1.76 "FTE Rate" shall mean, on a country-by-country or Region-by-Region basis in the Territory, in respect of performance of activities related to any Product, the applicable rate or rates with respect to the applicable FTEs in such country or Region based upon the fully-burdened cost of such FTE as determined pursuant to Section 6.9 (FTE Rates). The FTE Rate shall be inclusive of [***], including [***], such as [***] in each case, [***], but shall exclude [***]. The FTE Rate shall include the Field Force FTE Rates.

1.77 "Fully-Burdened Costs" shall mean, with respect to any activity under this Agreement, the sum of Out-of-Pocket Costs and FTE Costs for such activity.

1.78 "GAAP" shall mean generally accepted accounting principles, as applicable in the United States.

1.79 "Good Practices" shall mean compliance with the applicable standards contained in then-current "Good Laboratory Practices," or "GLP" "Good Manufacturing Practices" or "GMP" or "Good Clinical Practices" or "GCP" as promulgated by the FDA, and all analogous guidelines promulgated by the EMA or the ICH, as applicable.

1.80 "Governmental Authority" shall mean any court, agency, authority, council, commission, tribunal, department, regulatory body, or other instrumentality of any government or country or of any multi-national, national, federal, state, provincial, regional, county, municipal, city, or other political subdivision of any such government or any supranational organization of which any such country is a member.

1.81 "IND" shall mean an Investigational New Drug Application filed with the FDA pursuant to 21 C.F.R.§ 312, including all amendments and supplements to such application, or any equivalent filing with any Regulatory Authority outside of the United States.

1.82 "Indemnified Party" has the meaning set forth in Section 14.3.1 (Notice).

1.83 "Indemnifying Party" has the meaning set forth in Section 14.3.1 (Notice).

1.84 "Indication" shall mean any use of a Product for the treatment, prevention, cure, or to delay the progression of a human disease or condition.

1.85 "Infringement Claim" has the meaning set forth in Section 10.11.1 (Notice of Infringement Claims).

1.86 "Initial Development Plan" has the meaning set forth in Section 5.4 (Development Plan).

1.87 "Intellectual Property Rights" shall mean any Know-How, Patent Rights, copyrights, trade secrets, and any other intellectual property rights throughout the world, excluding Trademarks.

1.88 "Joint IP" shall mean all Joint Know-How and Joint Patent Rights, but excluding Product IP, Kiniksa Platform IP, and Kiniksa Product Data.

1.89 "Joint Know-How" shall mean all Know-How, invented, discovered, conceived, created, reduced to practice, or otherwise generated jointly by the Parties or their Affiliates in the performance of activities under this Agreement, but excluding all Kiniksa Product Data, Product IP, or Kiniksa Platform IP.

1.90 "Joint Patent Rights" shall mean any Patent Rights that include at least one claim that Covers Joint Know-How.

1.91 "Joint Steering Committee" or "JSC" has the meaning set forth in Section 4.1.1 (Formation, Composition, and Membership).

1.92 "Kiniksa Corp" has the meaning set forth in Section 17.10 (Guarantee).

1.93 "Kiniksa Field" shall mean all Indications and routes of administration, excluding (a) the Regeneron Retained EEO Field, (b) the Regeneron Field, until the date of receipt of U.S. Marketing Approval (and upon such date of receipt of U.S. Marketing Approval, the Kiniksa Field shall automatically expand to include the Regeneron Field in accordance with Section 2.1.4 (Kiniksa Field; Territory)), and (c) the Retained Field; *provided that* the Kiniksa Field may be expanded in accordance with Section 2.1.4 (Kiniksa Field; Territory) and Section 2.6 ([***]).

1.94 "Kiniksa Field Positive Clinical Readout" shall mean the [***] as set forth in the Development Plan that indicates (a) [***] and (b) an [***].

1.95 "Kiniksa Indemnitees" has the meaning set forth in Section 14.2 (Indemnity by Regeneron).

1.96 "Kiniksa Platform IP" shall mean all Intellectual Property Rights that are invented, discovered, conceived, created, reduced to practice, or otherwise generated solely (*i.e.*, without Regeneron) by or on behalf of Kiniksa or its Affiliates in the performance of activities under this Agreement that are (a) necessary to Develop, Manufacture, or Commercialize any Product in the Kiniksa Field in the Territory, or (b) useful to Develop, Manufacture, or Commercialize any Product in the Kiniksa Field in the Territory and actually used during the Term by Kiniksa or its Affiliates in connection therewith, in each case, excluding any Product IP and Kiniksa Product Data.

1.97 "Kiniksa Product Data" shall mean clinical data or Commercial Information primarily related to the Product for any Indication in the Kiniksa Field in the Territory generated by or on behalf of Kiniksa or its Affiliates.

1.98 "Know-How" shall mean any and all proprietary technical, scientific, or other information, data (including physical, chemical, biological, toxicological, pharmacological, clinical and veterinary data), test results, knowledge, know-how, techniques, practices, discoveries, inventions, specifications, dosage regimens, control assays, product specifications, analytical and quality control data, marketing, pricing, distribution cost and sales data or descriptions, designs, trade secrets, Regulatory Documentation, and other technology, whether or not written or otherwise fixed in any form or medium, regardless of the media on which contained and whether or not patentable or otherwise protected by trade secret law, in each case, that is not in the public domain or otherwise generally known.

1.99 "Knowledge" shall mean, for each Party, the actual knowledge of such Party's internal legal department (including such Party's intellectual property group), any employees of

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such Party who were directly involved in the preparation of this Agreement with the other Party.

1.100 "Launch Preparation Expenses" shall mean, with respect to a Product, on a country-by-country basis in the Territory, all expenses incurred prior to receipt of Marketing Approval for such Product in such country in connection with any Commercialization activities.

1.101 "Legal Dispute" shall mean any dispute related to a Party's alleged failure to comply with this Agreement or the validity, breach, termination, or interpretation of this Agreement.

1.102 "Lonza" shall mean Lonza Biologics PLC.

1.103 "Lonza License" shall mean that certain License Agreement dated December 1, 2000 by and between Lonza and Regeneron, as amended by Amendment No.1, dated August 10, 2016.

1.104 "Major Market Country" shall mean any of the following: (i) [***] and (ii) [***].

1.105 "Manufacture" or "Manufacturing" shall mean activities directed to producing, manufacturing, processing, packaging, labeling, devices and other delivery technologies, filling, finishing, assembly, quality assurance, quality control, testing, and release, shipping, or storage of any Product (or any components or process steps involving any Product), placebo, or comparator agent, as the case may be, including process development, qualification, and validation, scale-up, pre-clinical, clinical, and commercial manufacture and analytic development, product characterization, and stability testing.

1.106 "Manufacturing Cost" means the [***] if such Product (or any component thereof) is Manufactured by a Third Party, the amounts paid by a Party to such Third Party for Products as invoiced to such Party without any mark-up, including all Costs of Finishing (and will include any fees paid to such Third Party [***] to the extent not already included in the Manufacturing Cost), in each case, excluding all costs and expenses incurred by or on behalf either Party in connection with [***]. Manufacturing Costs shall include costs incurred with respect to, or as a result of, [***]. Manufacturing Costs shall include depreciation related to [***] incurred by either Party in providing [***] the Manufacture of Products, to be calculated using methodology that is in accordance with GAAP and consistently applied by such Party throughout its operations (but for clarity, not the [***]).

If a Product is Manufactured in a facility that is also used to manufacture other products not included within the scope of this Agreement, then only the value of the specific resources used for or reasonably allocated to the Manufacture of Products shall be included in the calculation of Manufacturing Cost, and Manufacturing Costs will not include the cost of idle or underutilized capacity in such a shared facility.

1.107 "[***]" has the meaning set forth in [***].

1.108 "Manufacturing Direct Costs" equals the sum of the following to the extent incurred in connection with any Product:

(a) Direct labor costs, based on [***]; and

(b) Materials and supplies costs for making Products, based on actual costs, including any applicable [***].

1.109 "Manufacturing Indirect Costs" equals the sum of the following to the extent incurred in connection with Products:

(a) Costs for [***], in each case, shall be allocated to the cost of Products based on the proportion of [***]; and

(b) Overhead costs required to support the Manufacture of Products, which costs shall be [***]. Overhead costs allocated based on [***]. Overhead costs allocated based on [***].

1.110 "Manufacturing Technology Transfer" has the meaning set forth in Section 8.15.1 (Transfers).

1.111 "Manufacturing Technology Transfer Event" has the meaning set forth in Section 8.14 (Manufacturing Technology Transfer Event).

1.112 "Manufacturing Technology Transfer Costs" shall mean the Fully-Burdened Costs of each Party incurred in performing (or having performed) a Manufacturing Technology Transfer, but excluding Kiniksa's Fully-Burdened Costs incurred in connection with a Fill/Finish Technology Transfer undertaken in accordance with Section 8.14.2 (Fill/Finish Transfer) (for clarity, without an accompanying Formulated Bulk Technology Transfer).

1.113 "Manufacturing Technology Transfer Cost Balance" has the meaning set forth in Section 9.4.7(b) (Manufacturing Technology Transfer Cost Balance).

1.114 "Marketing Approval" shall mean any approval, registration, license, or authorization from the applicable Regulatory Authority that is necessary for the marketing and sale of a pharmaceutical product in a country or other regulatory jurisdiction in the Territory, excluding any separate Pricing Approval.

1.115 "Modified Clause" has the meaning set forth in Section 17.8 (Severability).

1.116 "Neovii" means Neovii Pharmaceuticals AG.

1.117 "Neovii License Agreement" shall mean that certain License and Supply Agreement, dated as of August 17, 2016, by and between Regeneron and Neovii.

1.118 "Net Sales" shall mean, with respect to a Product, the gross amount invoiced for *bona fide* arms' length sales of such Product in the Territory by or on behalf of Kiniksa or its Affiliates to Third Parties (including licensees, sublicensees, or distributors (but not wholesalers)), but excluding [***] less the following deductions, in each case, determined in accordance with GAAP consistently applied:

(a) normal and customary trade, cash, quantity and free-goods allowances granted and taken directly with respect to sales of such Product;

(b) amounts repaid or credited with respect to such Product by reason of defects, rejections, recalls, returns, rebates, or allowances;

(c) chargebacks, rebates, discounts, and other amounts paid on sale or dispensing of such Product;

(d) Third Party cash rebates and chargebacks related to sales of such Product, to the extent allowed;

(e) retroactive price reductions for such Product that are actually allowed or granted;

(f) compulsory payments, refunds, credits, and rebates directly related to the sale of such Product, accrued, paid, or deducted pursuant to agreements with government entities or payors (including managed care agreements) or governmental regulations, including government levied fees as a result of healthcare reform policies;

(g) [***];

(h) freight, postage, shipment, and insurance costs and customs duties incurred in delivering such Product that are separately identified on the invoice or other appropriate documentation;

(i) sales taxes, excess duties, or other consumption taxes and compulsory payments to Governmental Authorities related to the sale of such Product (but specifically excluding income tax), which taxes and duties are separately identified on the invoice;

(j) if and to the extent expressly agreed in writing by the Parties, any other specifically identifiable costs or charges included in the gross invoiced sales price of such Product falling within categories substantially equivalent to those listed above and ultimately credited to customers or a Governmental Authority or agency thereof.

Net Sales in currency other than United States Dollars shall be translated into United States Dollars according to the provisions of Section 9.4.6 (Invoices and Documentation).

Sales of Product between Kiniksa and its Affiliates for resale shall be disregarded for purposes of calculating Net Sales. Any of the items set forth above that would otherwise be deducted from the invoice price in the calculation of Net Sales, but that are separately charged to and paid by Third Parties, shall not be deducted from the invoice price in the calculation of Net Sales.

In the case of any sale of a Product for consideration other than cash, such as barter or countertrade, then (i) the gross amount invoiced for such Product for purposes of calculating Net Sales of such Product shall equal the weighted average invoiced sales price of such Product sold in the same country during the same reporting period, or (ii) if no such invoiced sales occur in such country in such reporting period, then the Net Sales of such Product shall equal the fair market value of the consideration received as reasonably agreed by the Parties. For clarity, the foregoing shall not apply to Product provided free of charge and no consideration, for research, testing, clinical trials, or other humanitarian purposes, including expanded access programs, patient assistance programs, or charitable donations.

1.119 "New Product Trademarks" shall mean the Trademarks Controlled by Kiniksa during the Term for use with any Product.

1.120 "Non-Conforming Product" shall mean, with respect to any Product, (a) any failure to conform to the Product Specifications set forth in the applicable Supply Agreement or Quality Agreement, or (b) that the Product was not manufactured in accordance with cGMP and Applicable Law.

1.121 "Non-Responsible Party" means the Party that is not the Responsible Party.

1.122 "OFAC" means the Office of Foreign Assets Control of the United States Department of the Treasury or any successor agency thereto.

1.123 "Other Shared Expenses" shall mean (a) those costs and expenses [***], (b) of all costs and expenses incurred in connection with [***], and (c) other costs or expenses agreed in writing by the Parties to be included as Other Shared Expenses, in each case, excluding any such costs to the extent that [***]. Other Shared Expenses exclude all costs and expenses included in Shared Commercial Expenses and COGS.

1.124 "Out-of-Pocket Costs" shall mean costs and expenses paid to Third Parties (or payable to Third Parties and accrued in accordance with GAAP) by Regeneron (or its Affiliate) or Kiniksa (or its Affiliate) for the performance of activities under this Agreement (including in connection with the Development, Manufacture, or Commercialization of a Product).

1.125 "Patent Application" shall mean any application for a Patent, including any provisional, non-provisional, continuation, continuation-in-part, or divisional application, and any PCT international application or national phase application, whether in the U.S. or in any foreign country.

1.126 "Patent Rights" shall mean Patents and Patent Applications, and without limiting the foregoing, the right to claim priority to such Patents and Patent Applications.

1.127 "Patents" shall mean any patent (including any reissue, extension, substitution, confirmation, re-registration, re-examination, revival, supplementary protection certificate, or patents of addition), whether in the U.S. or in any foreign country.

1.128 "Payee Party" has the meaning set forth in Section 9.8 (Taxes).

1.129 "Paying Party" has the meaning set forth in Section 9.8 (Taxes).

1.130 "Payments" has the meaning set forth in Section 9.8 (Taxes).

1.131 "Person" shall mean an individual, partnership, limited liability partnership, joint venture, limited liability company, joint stock company, corporation, firm, trust, unincorporated organization, or other similar entity or organization, including a government or political subdivision or other department or agency thereof.

1.132 "Pharmacovigilance Agreement" has the meaning set forth in Section 7.6 (Pharmacovigilance).

1.133 "Phase III Trial" shall mean a human clinical trial that would satisfy the requirements of 21 C.F.R.§ 312.21(c) (as amended or any replacement thereof), including a human clinical trial that becomes a registration trial sufficient for filing a Marketing Approval, or an equivalent clinical trial in a country other than the U.S.

1.134 "Pricing Approval" shall mean such approval, agreement, determination, or decision establishing prices that can be charged to consumers for a pharmaceutical product or that will be reimbursed by Governmental Authorities for a pharmaceutical product, in each case, in a country where Governmental Authorities or Regulatory Authorities of such country approve or determine pricing for pharmaceutical products for reimbursement or otherwise.

1.135 "Product" shall mean any pharmaceutical product, drug product, preparation, formulation, or dosage form thereof that has rilonacept [***], including the pharmaceutical product for human use, containing rilonacept and developed or commercialized by Regeneron in the U.S. (known as ARCALYST ® (rilonacept) Injection for Subcutaneous Use in the United States) in the Existing Product Formulation. Notwithstanding the foregoing, for purposes of Section 1.138 (Product Manufacturing Records), Section 8.2 (Development Supply), Section 8.3 (Development Supply Agreement), Section 8.4 (Commercial Supply), Section 8.5 (Commercial Supply Agreement), Section 8.8 (Product Changes), Section 8.13 (Notification and Discussion of Supply Issues), and Section 8.14.4 (Failure to Supply), [***].

1.136 "Product Changes" shall mean changes to the dose, formulation, delivery, or form of any Product, or other changes to the method of Manufacturing of any Product for

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Commercialization in the Kiniksa Field in the Territory as compared to the dose, formulation, delivery, or form of such Product in the Existing Product Formulation, or other changes to the method of Manufacturing agreed by the Parties under any Supply Agreement or Quality Agreement.

1.137 "Product IP" shall mean all inventions and Intellectual Property Rights thereto that are invented, discovered, conceived, created, reduced to practice, or otherwise generated by a Party or its Affiliates (or jointly by the Parties or their Affiliates) in the performance of activities under this Agreement and that are exclusively or primarily related to a Product (and not generally related or generally applicable to pharmaceutical products), but excluding Kiniksa Product Data.

1.138 "Product Manufacturing Records" shall mean, with respect to a Product, copies of the files, documents, instruments, papers, books, and records Controlled by Regeneron or its Affiliates, whether in electronic or tangible form, including tangible embodiments of Regeneron Know-How, sufficient to enable a Person skilled in the art to Manufacture such Product (but for clarity, excluding Manufacturing the Product solely for the Retained Field or in the Retained Territory). For clarity, the foregoing shall include analytical and quality control data and results, cell culture methods and media compositions, formulae, and process and materials (excluding Product components and raw materials thereof) such as compositions of matter, cells, cell lines (excluding the master cell bank), assays, animal models and other physical, biological, or chemical material.

1.139 "Product Records" shall mean, with respect to a Product, copies of the files, documents, instruments, papers, books and records Controlled by Regeneron or its Affiliates, whether in electronic or tangible form, containing the administrative, safety, efficacy, quality, non-clinical and clinical data, regulatory, technical, chemistry and manufacturing control data (a) in the U.S. BLA; (b) that was submitted to EMA and maintained by Regeneron until the withdrawal by Regeneron of the BLA submitted to the EMA; (c) in any other Regulatory Documentation or other documents, reports, or information submitted to or correspondence with the FDA or any Governmental Authority related to such Product in the Territory (but excluding those specifically relating to the Retained Field or the Regeneron Retained EEO Field) and in Regeneron's possession or that may be readily available to Regeneron, or (d) in additional regulatory information regarding such Product that Kiniksa may request from Regeneron and in Regeneron's possession or that may be readily available to Regeneron. The Product Records exclude the Product Manufacturing Records.

1.140 "Product Specifications" shall mean the specifications for a Product, which specifications shall be set forth in the Quality Agreements.

1.141 "Product Trademarks" shall mean, with respect to the Product in the Kiniksa Field in the Territory, the Existing Product Trademarks and the New Product Trademarks.

1.142 "Profit Payment Report" shall mean the consolidated Quarterly report prepared by Kiniksa (based on information reported under Section 9.4.1 (Periodic Reports)) setting forth in reasonable detail in the aggregate for the Territory as a whole (a) the Net Sales invoiced by Kiniksa or its Affiliates during such Quarter, and (b) the Other Shared Expenses, Cost of Goods Sold, and Shared Commercial Expenses, in each case, incurred by each Party for such Quarter.

1.143 "Profit Split Arrangement" shall mean the sublicense revenue sharing arrangement between the Parties for the Product as described in Section 9.3 (Sharing of Third Party Proceeds) and the profit-sharing arrangement between the Parties for the Product described in Section 9.4 (Sharing of Profits).

1.144 "Profits" shall mean, for the Product during a given period of time, (a) the Net Sales invoiced by Kiniksa or its Affiliates during such period, *less* (b) the sum of (i) Cost of Goods Sold for Product used, sold, otherwise distributed for patient use, or that expires during such period, (ii) Shared Commercial Expenses incurred by either Party during such period attributable to such Product, (iii) Other Shared Expenses incurred by either Party during such period attributable to such Product, (iv) Manufacturing Technology Transfer Costs, and (v) the Manufacturing Technology Transfer Cost Balance, to the extent permitted in accordance with Section 9.4.7(b) (No Carryforward, Exception).

1.145 "Projected Net Sales" has the meaning set forth in Section 9.5.2 (Promotion Exclusive Negotiation Period).

1.146 "Promotion" shall mean the marketing, detailing, advertising, or other promotion of a Product, but excluding the sale of such Product.

1.147 "Promotion Exclusive Negotiation Period" has the meaning set forth in Section 2.10.3 (Promotion Exclusive Negotiation Period).

1.148 "Promotion ROFN" has the meaning set forth in Section 2.10.1 (Promotion ROFN).

1.149 "Promotion ROFN Exercise Notice" has the meaning set forth in Section 2.10.2 (Promotion ROFN Exercise Notice).

1.150 "Promotion ROFN Notice" has the meaning set forth in Section 2.10.1 (Promotion ROFN).

1.151 "Public Official or Entity" shall mean (a) any officer, employee, agent, representative, department, agency, de facto official, corporate entity, or instrumentality or subdivision of any government, military, or international organization, including any state-owned or affiliated company or hospital, or (b) any candidate for political office, any political party, or any official of a political party.

1.152 "Quality Agreements" has the meaning set forth in Section 8.6 (Quality Agreements).

1.153 "Quarter" or "Quarterly" shall refer to a calendar quarter, except that the first (1st) Quarter of the Term shall commence on the Effective Date and extend to the end of the then-current calendar quarter, and the last calendar quarter of the Term shall extend from the first day of such calendar quarter until the effective date of the termination or expiration of this Agreement.

1.154 "Quarterly Expense Report" has the meaning set forth in Section 9.4.1(b) (Quarterly Incurred Expenses).

1.155 "Recall Responsible Party" has the meaning set forth in Section 7.8 (Recalls and Other Corrective Actions).

1.156 "Receiving Party" has the meaning set forth in Section 13.1 (Confidential Information).

1.157 "Regeneron Exploratory Clinical Studies" has the meaning set forth in Section 3.6 **(**Investigator Initiated Studies and Regeneron Exploratory Clinical Studies).

1.158 "Regeneron Field" shall mean, until the date of receipt of U.S. Marketing Approval, CAPS and DIRA, in each case, in the United States and Japan. After the date of receipt of U.S. Marketing Approval, the Regeneron Field will be included in the Kiniksa Field.

1.159 "Regeneron Indemnitees" has the meaning set forth in Section 14.1 (Indemnity by Kiniksa).

1.160 "Regeneron IP" shall mean the Regeneron Patent Rights and the Regeneron Know-How. For clarity, [***].

1.161 "Regeneron Know-How" shall mean all Know-How Controlled by Regeneron or its Affiliates as of the Effective Date or during the Term that (a) is necessary to Develop or Commercialize a Product in the Kiniksa Field in the Territory or to Manufacture a Product, (b) is useful to Develop or Commercialize a Product in the Kiniksa Field in the Territory, or Manufacture a Product and is actually used by or on behalf of Regeneron or its Affiliates in connection with the Development, Commercialization, or Manufacture of any Product, or (c) constitutes Know-How included in the Product IP. Notwithstanding the foregoing, Regeneron Know-How shall not include any [***] and any other [***] for Kiniksa pursuant to Section 5.6 (Regeneron Performance of Sample Bioanalysis). For clarity, the Regeneron Know-How includes the Regeneron Manufacturing Know-How and excludes Joint Know-How.

1.162 "Regeneron Manufacturing IP" shall mean the Regeneron Manufacturing Patent Rights and the Regeneron Manufacturing Know-How.

1.163 "Regeneron Manufacturing Know-How" shall mean all Know-How Controlled by Regeneron or its Affiliates as of the Effective Date or during the Term that is (a) necessary to Manufacture any Product, or (b) useful to Manufacture a Product and is

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actually used by or on behalf of Regeneron or its Affiliates in connection with such Manufacture.

1.164 "Regeneron Manufacturing Patent Rights" shall mean the Regeneron Patent Rights that include at least one (1) claim that specifically Covers the Manufacture of a Product (but that does not include any claims that otherwise Cover the Development or Commercialization of such Product).

1.165 "Regeneron Patent Rights" shall mean all Patent Rights Controlled by Regeneron or its Affiliates as of the Effective Date or during the Term that includes at least one (1) claim that Covers the Development or Commercialization of a Product in the Kiniksa Field in the Territory or specifically Covers the Manufacture of a Product. The Regeneron Patent Rights as of the Effective Date are set forth on Schedule 1.165 (Regeneron Patent Rights). For clarity, the Regeneron Patent Rights include the Regeneron Manufacturing Patent Rights [***] and exclude Joint Patent Rights and the Patent Rights within the Kiniksa Platform IP.

1.166 "Regeneron Profit Percentage" shall mean during a given period of time, fifty percent (50%) of the Profits for such Product during such period.

1.167 "Regeneron Profit Split" shall mean, during a given period of time, the product of (a) Profits in such period, and (b) the Regeneron Profit Percentage.

1.168 "Regeneron Profit Split Payment" has the meaning set forth in Section 9.4.5(c) (Payment to Regeneron).

1.169 "Regeneron Retained EEO Field" shall mean (a) local administration to the eye for any Indication, (b) local administration to the ear for any Indication, and (c) any Indication in oncology.

1.170 "Regeneron Third Party Proceeds Percentage" shall mean during a given period of time, fifty percent (50%) of the Third Party Proceeds actually received by Kiniksa or its Affiliates during such period, on a cash basis.

1.171 "Regeneron Third Party Proceeds Split" shall mean, during a given period of time, the product of (a) Third Party Proceeds actually received by Kiniksa or its Affiliates during such period, on a cash basis, and (b) the Regeneron Third Party Proceeds Profit Percentage during such period.

1.172 "Regeneron Third Party Proceeds Split Payment" has the meaning set forth in Section 9.4.5(d) (Payment to Regeneron of the Regeneron Third Party Proceeds Payment).

1.173 "Region" shall mean a group of countries as approved by the JSC pursuant to Section 4.1.3(a) (Duties of the JSC).

1.174 "Registration Filing" shall mean the submission to the relevant Regulatory Authority of an appropriate application seeking Approval, and shall include any IND, BLA, or other application for Marketing Approval.

1.175 "Regulatory Authority" shall mean any federal, national, multinational, state, regional, provincial, or local regulatory agency, department, bureau, or other governmental entity anywhere in the world with authority with respect to development, manufacturing, marketing, commercialization, reimbursement, or pricing of a pharmaceutical product, including the FDA in the United States.

1.176 "Regulatory Documentation" shall mean all (a) regulatory applications, submissions, dossiers, notifications, registrations, Registration Filings, Approvals, or other filings made to or with, or other approvals granted by, a Regulatory Authority or other Governmental Authority that are necessary or reasonably desirable in order to Develop, Manufacture, or Commercialize a Product in a particular country or regulatory jurisdiction in the Territory, (b) correspondence and reports submitted to or received from any Regulatory Authority or other Governmental Authority in the Territory (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 (c) clinical and other data contained or relied upon in any of the foregoing; in each case ((a), (b), and (c)), relating to a Product.

1.177 "Regulatory Materials" has the meaning set forth in Section 7.3 (Certain Regulatory Materials).

1.178 "Responsible Party" means, unless otherwise expressly set forth in this Agreement, Regeneron prior to the date of receipt of U.S. Marketing Approval, and Kiniksa on and after the date of receipt of U.S. Marketing Approval.

1.179 "Retained Field" shall mean the Indications set forth on Schedule 1.179, in each case, outside of the United States and Japan, but otherwise inside of the Territory; [***]**]**.

1.180 "[***]" has the meaning set forth in [***].

1.181 "Retained Territory" shall mean the countries and territories set forth on Schedule 1.181.

1.182 "Safety Termination" has the meaning set forth in Section 16.5 (Termination by Kiniksa).

1.183 "Sale Agreement" has the meaning set forth in Section 2.11.1 (Sale ROFN).

1.184 "Sale Exclusive Negotiation Period" has the meaning set forth in Section 2.11.3 (Sale Exclusive Negotiation Period).

1.185 "Sale Negotiations" has the meaning set forth in Section 2.11.1 (Sale ROFN).

1.186 "Sale ROFN" has the meaning set forth in Section 2.11.1 (Sale ROFN).

1.187 "Sale ROFN Exercise Notice" has the meaning set forth in Section 2.11.2 (Sale ROFN Exercise Notice).

1.188 "Sale ROFN Notice" has the meaning set forth in Section 2.11.1 (Sale ROFN).

1.189 "Shared Commercial Expenses" shall mean the sum of the following items, in each case, on a country-by-country basis, incurred after Marketing Approval for any Product in a country in the Territory, including U.S. Marketing Approval with respect to the United States, to the extent directly attributable to Commercialization of the Product in the Kiniksa Field in the Territory in accordance with the Commercial Plan and to the extent that such items do not include any costs included in Manufacturing Costs, Other Shared Expenses, or COGS:

(a) Field Force Costs;

(b) Out-of-Pocket Costs for (i) the Promotion of Products in the Kiniksa Field in the Territory (including pricing activities, educational expenses, advocate development programs and symposia, and promotional materials), but for clarity excluding costs for Detailing, which costs shall be covered under Field Force Costs, (ii) market research for Products, (iii) the preparation of training and communication materials for the Product, and (iv) Third Party contractors engaged by Kiniksa to Promote the Product;

(c) (i) Out-of-Pocket Costs for surveys, registries, [***] for the Product, including Out-of-Pocket Costs paid to clinical research organizations or incurred as investigator and expert fees, lab fees, or scientific service fees, and (ii) Out-of-Pocket Costs of shipping clinical supplies to centers or disposal of clinical supplies for registries [***], in each case ((i) and (ii)), to the extent not already included in the Cost of Goods Sold for such Product;

(d) Out-of-Pocket Costs incurred in connection with obtaining Pricing Approvals and the maintenance of all Marketing Approvals, in each case, for the Product;

(e) Out-of-Pocket Costs related to (i) Development activities performed to maintain Marketing Approval (but not including Development activities required by a Regulatory Authority in any country in the Territory as a condition of obtaining Marketing Approval in the Kiniksa Field in such country), and (ii) regulatory affairs activities performed to maintain Marketing Approval (but not including regulatory affairs activities required to obtain Approval of additional labeled Indications, line extensions, or formulations of a Product in the Kiniksa Field in any country in the Territory);

(f) Any Manufacturing Direct Costs or Manufacturing Indirect Costs incurred by or on behalf of a Party to supervise or coordinate any Manufacturing of Products performed by the other Party or any Third Party;

(g) Commercial Overhead Charge for each country or Region;

(h) bad debt attributable to sales of the Product in the Territory that have been written off by Kiniksa or its Affiliates;

(i) [***] of Net Sales of the Product to cover the cost of distribution and warehousing for the sale of the Product in the Territory, to the extent not already deducted from Net Sales of the Product pursuant to clause 1.118(h) of the definition of Net Sales for such Product (and which for clarity shall be a proxy for costs actually incurred) or otherwise included in Manufacturing Costs;

(j) Third Party License Payments to the extent attributable to the Commercialization of a Product; *provided that* [***], then the [***] that are [***]; and

(k) any other FTE Costs or Out-of-Pocket Costs (i) related to the Commercialization of any Product and not included in clauses 1.189(a) through 1.189(j) above, and specifically identified and included in the Commercial Plan that are reasonable and customary Commercialization expenses, or (ii) included as Shared Commercial Expenses by agreement of the Parties.

For clarity, Shared Commercial Expenses shall not include Launch Preparation Expenses. In the event that any of the costs included in Section 1.189(a) through Section 1.189(k) [***], then [***]. In no event shall the same costs be included more than once in Shared Commercial Expenses under this Agreement.

1.190 "Subsequent Indication [***]" has the meaning set forth in Section 9.5.3 ([***]).

1.191 "Subsequent Indication Launch Plan" has the meaning set forth in Section 9.5.3 ([***]).

1.192 "Supply Agreements" has the meaning set forth in Section 8.5 (Commercial Supply Agreement).

1.193 "Technical Consultation and Transition Services" has the meaning set forth in Section 3.10.1 (Provision of Services).

1.194 "Technology Transfer Plan" has the meaning set forth in Section 8.15.1 (Transfers).

1.195 "Term" has the meaning set forth in Section 16.1 (Term).

1.196 "Territory" shall mean all the countries and territories of the world, other than the Retained Territory.

1.197 "Third Party" shall mean any Person other than Kiniksa, Regeneron, or any Affiliate of either Party.

1.198 "Third Party Fill/Finish Provider" shall mean any Third Party engaged to perform filling, finishing, packaging, or labeling of a Product.

1.199 "Third Party License Payment" shall mean any payment due to any Third Party under any Third Party License, including upfront payments, royalties, and milestone payments.

1.200 "Third Party Licenses" shall mean any agreement between a Party and a Third Party (a) pursuant to which such Third Party grants a license to such Party with respect to Intellectual Property Rights of such Third Party in order to (i) avoid an allegation of infringement or misappropriation of any Third Party's Patent Rights, Know-How, or other Intellectual Property Rights in connection with the Development, Manufacture, or Commercialization of Products in such Party's territory, or (ii) obtain access to Patent Rights, Know-How or other Intellectual Property Rights that may be colorably necessary or useful to the Development, Manufacture, or Commercialization of Products in such Party's territory, and (b) into which the JSC approves the entry pursuant to Section 4.1.3(t) (Duties of the JSC). Third Party Licenses include the Existing Licenses.

1.201 "Third Party Offer" has the meaning set forth in Section 2.10.4 (Failure to Agree; Promotion Third Party Offers).

1.202 "Third Party Proceeds" shall mean, with respect to a Product, proceeds received by Kiniksa from any Third Party (including licensees, sublicensees and distributors) in consideration for the sale, license, or other disposition of rights with respect to such Product (including upfront payments, milestone payments, and royalties), but excluding payments received by Kiniksa or any of its Affiliates (a) in connection with a Change of Control of Kiniksa [***], (b) in connection with any equity investments at fair market value or loans made at market interest rates, or (c) reimbursement for patent expenses at Kiniksa's Out-of-Pocket Cost.

1.203 "Third Party Proceeds Payment Report" has the meaning set forth in Section 9.4.1(c) (Periodic Reports).

1.204 "Trademarks" shall mean all registered and unregistered trademarks (including all common law rights thereto), service marks, product marks, trade names, brand names, logos, taglines, slogans, certification marks, Internet domain names, trade dress, corporate names, business names, and other indicia of origin, together with the goodwill associated with any of the foregoing, and all applications, registrations, extensions, and renewals thereof throughout the world, and all rights therein provided by international treaties and conventions.

1.205 "United States" or "U.S." means the United States of America and its territories and possessions.

1.206 "Unresolved Matter" has the meaning set forth in Section 4.2.2(b) (Escalation).

1.207 "U.S. BLA" means BLA No.125249 registered with the FDA in the United States.

1.208 "U.S. BLA Transfer Date" means the date that the FDA accepts and confirms the transfer of the U.S. BLA from Regeneron to Kiniksa.

1.209 "U.S. Export Control Laws" shall mean all applicable U.S. laws and regulations relating to the export or re-export of commodities, technologies, or services, including the Export Administration Act of 1979, 24 U.S.C.§§ 2401-2420, the International Emergency Economic Powers Act, 50 U.S.C.§§ 1701-1706, the Trading with the Enemy Act, 50 U.S.C.§§ 1 et. seq., the Arms Export Control Act, 22 U.S.C.§§ 2778 and 2779, and the International Boycott Provisions of Section 999 of the U.S. Internal Revenue Code of 1986.

1.210 "U.S. Marketing Approval" shall mean the first Marketing Approval by the FDA of a Product for an Indication in the Kiniksa Field in the United States, but excluding any Approval for compassionate, named patient approval, or similar use.

1.211 "U.S. Promotion Negotiations" has the meaning set forth in Section 2.10.1 (Promotion ROFN).

1.212 "U.S. Third Party Promotion Agreement" has the meaning set forth in Section 2.10.2 (Promotion ROFN Exercise Notice).

1.213 "Valid Claim" shall mean either (a) a claim of an issued and unexpired Patent (including the term of any patent term extension, supplemental protection certificate, renewal or other extension) that has not been held unpatentable, invalid or unenforceable in a final decision of a court or other Governmental Authority of competent jurisdiction from which no appeal may be or has been taken, and that has not been admitted to be invalid or unenforceable through reissue, re-examination, disclaimer or otherwise, or (b) a pending claim of a Patent Application that was filed in good faith and has not been abandoned or finally disallowed without the possibility of appeal or refiling; *provided, however*, that Valid Claim will exclude any such pending claim in any such Patent Application that has not been granted within [***] after the earliest filing date from which such Patent Application takes priority.

1.214 "Working Group" has the meaning set forth in Section 4.1.1 (Formation, Composition, and Membership).

ARTICLE 2
LICENSES, RIGHT OF FIRST NEGOTIATION, AND NOTICE RIGHTS

2.1 **Regeneron Licenses to Kiniksa**.

2.1.1 **Exclusive License**. Subject to the terms of this Agreement, including Section 2.10 (Regeneron ROFN to Promote Products in the U.S.) and Section 3.6 **(**Investigator Initiated Studies and Regeneron Exploratory Clinical Studies), Regeneron shall grant and hereby grants to Kiniksa an exclusive, non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing)), license under (a) the Regeneron IP, other than the Regeneron Manufacturing IP, and (b) Regeneron's interest in the Joint IP, in each case ((a) and (b)), to (i) Develop the

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Product (whether inside or outside of the Territory) for Commercialization in the Kiniksa Field in the Territory, and (ii) Commercialize the Product in the Kiniksa Field in the Territory.

2.1.2 **Manufacturing License**. Subject to the terms of this Agreement, upon the occurrence of a Manufacturing Technology Transfer Event, Regeneron shall grant and hereby grants to Kiniksa a non-exclusive, non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing)) license under the Regeneron IP, including the Regeneron Manufacturing IP, and Regeneron's interest in the Joint IP, in each case, to Manufacture the Product for the uses set forth in clauses (b)(i) and (ii) of Section 2.1.1 (Exclusive License).

2.1.3 **Right of Reference**. Regeneron shall grant and hereby grants to Kiniksa a sublicensable right to cross-refer to all Approvals and Regulatory Documentation for the Product that are Controlled by Regeneron for use in the Kiniksa Field in the Territory; *provided that* [***].

2.1.4 **Kiniksa Field; Territory**. Upon U.S. Marketing Approval, the Kiniksa Field shall automatically expand to include the Regeneron Field for purposes of this Agreement. [***].

2.1.5 **Regeneron EEO Reserved Field and Regeneron Exploratory Clinical Studies**. Regeneron retains the right to (a) Develop and Commercialize the Product in the Regeneron EEO Reserved Field (whether inside or outside the Territory) and (b) perform Regeneron Exploratory Clinical Studies for the Product in the Kiniksa Field in accordance with Section 3.6 (Investigator Initiated Studies and Regeneron Exploratory Clinical Studies) (whether inside or outside the Territory).

2.2 **Kiniksa Licenses to Regeneron**.

2.2.1 **Exclusive License**. Subject to the terms of this Agreement, Kiniksa shall grant and hereby grants to Regeneron an exclusive (even as to Kiniksa and its Affiliates), non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing)), license under the Kiniksa Product Data and Kiniksa's interest in the Joint IP to (a) Develop the Product for Commercialization outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory (whether inside or outside of the Kiniksa Field), (b) Commercialize the Product outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory (whether inside or outside of the Kiniksa Field), and (c) Manufacture the Product for the uses set forth in clauses (a) and (b) of this Section 2.2.1 (Exclusive License).

2.2.2 **Non-Exclusive License**. Subject to the terms of this Agreement, Kiniksa shall grant and hereby grants to Regeneron a non-exclusive, non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable in multiple tiers (in accordance with Section 2.7 (Sublicensing)) license under the Kiniksa Platform IP to (a) Develop the Product for Commercialization outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory (whether inside or outside of the Kiniksa Field), (b) Commercialize the Product outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory (whether inside or outside of the Kiniksa Field), (c) Manufacture the Product for the uses set forth in clauses (a) and (b) of this Section 2.2.2 (Non-Exclusive License), and (d) to Manufacture the Product for Kiniksa for the uses set forth in clauses (b)(i) and (ii) of Section 2.1.1 (Exclusive License).

2.2.3 **Right of Reference**. Kiniksa shall grant and hereby grants to Regeneron a sublicensable right to cross-refer to all Approvals and Regulatory Documentation for the Product Controlled by Kiniksa for use (a) with the Product outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory (whether inside or outside of the Kiniksa Field), (b) to Manufacture the Product in accordance with the terms of this Agreement, and (c) in connection with development, commercialization or manufacture of all other products being developed or commercialized by Regeneron other than the Product. For clarity, Kiniksa shall grant and hereby grants to Regeneron a sublicensable right to cross-refer to all Approvals transferred by Regeneron to Kiniksa pursuant to Section 3.3.2 (Regulatory and Safety Information) for use by Regeneron in the Regeneron Field until U.S. Marketing Approval.

2.2.4 **Right of Reference for Purposes of Prosecution and Maintenance of Patent Rights**. Kiniksa shall grant and hereby grants to Regeneron a right to refer to the Kiniksa Product Data for purposes of prosecuting and maintaining Patent Rights within the Product IP.

2.3 **Right of Reference; Further Assurances**. The Party granting rights to the other Party under Section 2.1.3 (Right of Reference) or Section 2.2.3 (Right of Reference) (as applicable) will use reasonable efforts take such actions as may be reasonably requested by the other Party to give effect to the intent of such Sections and to give the other Party the benefit of the rights in such Sections. Such actions may include providing a signed statement that the other Party may rely on, and that the Regulatory Authority may access, in support of the other Party's application for Approval in its territory or providing information submitted by such Party to the Regulatory Authority with respect to any Registration Filing, Approval, or other Regulatory Documentation Controlled by such Party or its Affiliates that relates to any Product in each case, in a Party's possession or that may be readily available to such Party.

2.4 **Trademark License Grants to Kiniksa and Right to Use Regeneron Name and Logo**.

2.4.1 **Exclusive Trademark License**. Subject to the terms of this Agreement, Regeneron shall grant and hereby grants to Kiniksa an exclusive license sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing)) to use the Existing Product Trademarks in connection with the (a) Commercialization of the Product in the Kiniksa Field in the Territory, and (b) Development of the Product for purposes of Commercialization in the Kiniksa Field in the Territory.

2.4.2 **Non-Exclusive Trademark License**. Subject to the terms of this Agreement, upon the occurrence of a Manufacturing Technology Transfer Event, Regeneron shall grant and hereby grants to Kiniksa a non-exclusive, non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing)) license to use the Existing Product Trademarks in connection with the Manufacture of the Product for the uses set forth in clauses (b)(i) and (ii) of Section 2.1.1 (Exclusive License).

2.5 **Licenses Generally; No Implied License**. Except as expressly provided for herein, nothing in this Agreement grants either Party any rights, title, or interests in or to the Intellectual Property Rights, Trademarks, materials or Confidential Information of the other Party (either expressly or by implication or estoppel), and each Party specifically reserves all rights not expressly granted to the other Party. Regeneron retains the right to (a) Develop the Product for Commercialization outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory, and (b) Commercialize the Product outside of the Kiniksa Field (whether inside or outside of the Territory) and outside of the Territory. For clarity, Regeneron will not Develop the Product for Commercialization or Commercialize any Product, in each case, in the Kiniksa Field in the Territory.

2.6 [***]. The Parties [***] with respect to [***] for the [***] in each case, [***] Regeneron shall [***] related to [***].

2.7 **Sublicensing**. Each Party shall only enter sublicenses under the licenses granted in this Agreement in compliance with this Section 2.7 (Sublicensing) and the other applicable terms and conditions set forth in this Agreement. Each Party shall remain responsible and liable for the compliance by its sublicensees with applicable terms and conditions set forth in this Agreement. Any sublicense granted by a Party under this Section 2.7 (Sublicensing) will require the sublicensee to comply with the obligations of such Party contained herein, including the confidentiality and non-use obligations set forth in ARTICLE 13 (Confidentiality), and will include, with respect to a sublicense granted by Kiniksa, an obligation of the sublicensee to account for and report its sales of the Product to the sublicensing Party in a manner sufficient for such Party to comply with its reporting obligations under this Agreement. Notwithstanding anything to the contrary set forth in this Section 2.7 (Sublicensing), either Party may sublicense any of the rights

granted to it under this Agreement without the other Party's consent, (a) to an Affiliate, or (b) subject to Regeneron's Promotion ROFN as set forth in Section 2.10 (Regeneron ROFN to Promote Products in U.S.), to any Third Party subcontractor performing Development, Commercialization, or Manufacturing activities with respect to any Product on a fee-for-service basis (for clarity, without the right for such subcontractor to participate in future rights or revenues associated with the Product), as applicable, (e.g., contract research organization, contract manufacturing organization or contract sales organization) to the extent required for such Third Party to perform such services (including drug development or manufacturing services); provided that in the case of clause (b) that the subcontracting Party granting such sublicense has fully satisfied all of its obligations pursuant to Section 2.9 (Subcontracting), including any applicable subcontract consent requirements. Without limiting any consent rights in this Section 2.7 (Sublicensing), each Party will notify the other Party of any sublicense to its Affiliate or any Third Party no later than [***] days after the execution of such sublicense agreement. Kiniksa will forward to Regeneron a complete copy of each fully executed sublicense agreement (and any material amendments thereto) with a Third Party sublicensee no later than [***] days after the execution of such sublicense agreement.

2.7.1 **Sublicensing by Regeneron**. Regeneron may grant sublicenses under the licenses granted by Kiniksa in Section 2.2 (Kiniksa Licenses to Regeneron) [***].

2.7.2 **Sublicensing by Kiniksa**.

(a) Kiniksa may grant sublicenses under the licenses granted by Regeneron in Section 2.1.1 (Exclusive License), [***] Prior to [***] Kiniksa shall provide Regeneron with a summary of the financial and material terms of the proposed sublicense and a copy of the proposed sublicense agreement.

(b) Kiniksa may grant sublicenses under the licenses granted by Regeneron in Section 2.1.2 (Manufacturing License), [***]. For clarity, [***] shall apply to the granting of sublicenses in connection with fill/finish activities related to the Product, as provided in Section 8.1 (Regeneron Supply of Product).

(c) Kiniksa has the right to sublicense any of its rights under the licenses granted by Regeneron in Section 2.3 (Trademark License Grants to Kiniksa) to any permitted sublicensee of any Product.

2.7.3 **Continuation of Sublicenses Upon Termination**. If the licenses granted to Kiniksa pursuant to Section 2.1 (Regeneron Licenses to Kiniksa) or Section 2.4 (Trademark License Grants to Kiniksa) terminate as a result of a termination of this Agreement by Regeneron for Kiniksa's material breach pursuant to Section 16.3.1 (Material Breach) or for Kiniksa's insolvency pursuant to Section 16.4 (Termination for Insolvency), then, at the written request of any sublicensee

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who is not then in breach of the applicable sublicense agreement between Kiniksa and such sublicensee, Regeneron will negotiate in good faith to attempt to enter into a direct license agreement between Regeneron and such sublicensee under the Regeneron IP or Existing Product Trademarks (as applicable) sublicensed to such sublicensee of the same scope as set forth in such sublicense agreement between Kiniksa and such sublicensee; *provided*, *however*, that (a) such sublicensee undertakes to perform in all respects its obligations under the applicable sublicense agreement, (b) such direct license agreement would not impose on Regeneron any obligations over and above Regeneron's obligations to Kiniksa under this Agreement, and (c) the direct license agreement would require such sublicensee to pay Regeneron the same amount as the sublicensee would have paid to Kiniksa with respect to a sublicense of rights to the Product (had this Agreement survived) as a result of such sublicensee's performance under the sublicense agreement.

2.8 **Compliance with Third Party Licenses**. Except as contemplated by [***], each Party shall comply with all applicable terms and conditions of each Third Party License, and shall perform and take such actions as may be required to allow the Party that is the contracting party to such Third Party License to comply with its obligations thereunder, including obligations relating to sublicensing, patent matters, confidentiality, reporting, audit rights, indemnification, and diligence. Without limiting the foregoing, each Party shall prepare and deliver to the other Party any additional reports required under the applicable Third Party License, in each case, sufficiently in advance to enable the Party that is party to such Third Party License to comply with its obligations under the applicable Third Party License. Each Party agrees, upon the other Party's request, to provide the other Party with copies of any Third Party Licenses to which it is a party (subject to any confidentiality restrictions in any such Third Party License). Confidential Information of the providing Party or its counterparty appended or attached as schedules may be redacted from such copies, except to the extent that such information is required in order to enable the other Party to comply with its obligations to the providing Party under this Agreement with respect to such Third Party License.

2.8.1 **Payments Under Third Party Licenses**. Each Party shall be responsible for one hundred percent (100%) of any Third Party License Payments under the Third Party Licenses to which it is a party; *provided that* [***]. Except as contemplated by [***], if Regeneron breaches its payment obligation to a licensor under any Third Party License to which Regeneron is a party [***], then Kiniksa may (but will not be obligated to) make such payments directly to such licensor. In such event, Kiniksa may include such amounts as Shared Commercial Expenses hereunder.

2.8.2 **Breach or Termination of Third Party Licenses**. If (a) a Party receives notice of an alleged breach by such Party under a Third Party License to which it is a party, or (b) a Party intends to terminate a Third Party License to which it is a party, then, in each case ((a) and (b)), such Party will promptly, but in any event

no later than [***] days thereafter, provide written notice thereof to the other Party.

2.8.3 **Freedom to Obtain New Third Party Licenses**. For the avoidance of doubt, following the Effective Date, [***], each Party shall be free to enter into agreements between a Party and a Third Party pursuant to which such Third Party grants a license to such Party with respect to Intellectual Property Rights of such Third Party in order to avoid infringement or misappropriation of any Third Party's Patent Rights, Know-How, or other Intellectual Property Rights in such Party's territory or obtain access to Patent Rights, Know-How, or other Intellectual Property Rights that may be reasonably necessary or useful to the Development, Manufacture or Commercialization of Products in such Party's territory; *provided that* [***].

2.9 **Subcontracting**. Subject to compliance with Section 2.10 (Regeneron ROFN to Promote Products in the U.S.) and Section 2.7.2 (Sublicensing by Kiniksa), each Party may perform any of its obligations under this Agreement through one or more subcontractors; provided that (a) the subcontracting Party remains fully responsible for the work allocated to, and payment to, such subcontractors to the same extent it would if it had done such work itself; (b) the subcontractor undertakes in writing obligations of confidentiality and non-use regarding Confidential Information that are substantially the same as those undertaken by the Parties pursuant to ARTICLE 13 (Confidentiality); and (c) the subcontractor agrees in writing to assign all inventions and Intellectual Property Rights developed in the course of performing any such work under this Agreement to the Party retaining such subcontractor, or as otherwise required under this Agreement and upon request to sign any documents to confirm or perfect such assignment and to cooperate in the preparation and prosecution of any such inventions.

2.10 **Regeneron ROFN to Promote Products in the U.S**. Kiniksa may engage Third Party contractors to Promote the Product in the Kiniksa Field in the Territory; provided that any such contractor is either (a) [***], or (b) [***]. Except as permitted pursuant to the previous sentence, Kiniksa shall not enter into any negotiations with any Third Party pursuant to which Kiniksa would subcontract its rights to Promote any Product in the U.S. or execute any agreement granting any Third Party rights to the foregoing, except, in each case, in compliance with this Section 2.10 (Regeneron ROFN to Promote Products in the U.S.).

2.10.1 **Promotion ROFN**. Kiniksa hereby grants to Regeneron a right of first negotiation in the event Kiniksa wishes to enter a subcontracting arrangement to Promote any Product in the Kiniksa Field in the U.S. (such right, a "Promotion ROFN"). In such event, Kiniksa will deliver a notice to Regeneron of its desire to enter into such a subcontracting arrangement in the U.S. (the "Promotion ROFN Notice") prior to Kiniksa engaging in any negotiations with, accepting any offer from, or entering into any agreement, with any Third Party to

subcontract such Promotion rights (collectively, "U.S. Promotion Negotiations").

2.10.2 **Promotion ROFN Exercise Notice**. In the event Regeneron wishes to enter into exclusive U.S. Promotion Negotiations with Kiniksa to obtain the rights that Kiniksa wishes to grant with respect to the Promotion of any such Product in the Kiniksa Field in the U.S., Regeneron shall provide Kiniksa with notice thereof ("Promotion ROFN Exercise Notice") within [***] after Regeneron's receipt of the Promotion ROFN Notice. If Regeneron fails to deliver the Promotion ROFN Exercise Notice to Kiniksa within such [***] period, then Kiniksa shall thereafter be free to engage in U.S. Promotion Negotiations with Third Parties for, and enter into a written agreement with, any Third Party with respect to the Promotion of such Product (each a "U.S. Third Party Promotion Agreement") without further obligations under this Section 2.10 (Regeneron ROFN to Promote Products in the U.S.).

2.10.3 **Promotion Exclusive Negotiation Period**. In the event Regeneron delivers the Promotion ROFN Exercise Notice within the [***] time period set forth in Section 2.10.2 (Promotion ROFN Exercise Notice), the Parties will engage in good faith negotiations, and Kiniksa will permit Regeneron to conduct Regeneron's conduct of, technical due diligence for a period of [***] after delivery of the Promotion ROFN Exercise Notice ("Promotion Exclusive Negotiation Period") in an attempt to agree upon the terms and conditions pursuant to which Regeneron would receive a right to Promote such Product in the U.S. in the Kiniksa Field. If the Parties are able to reach agreement on such terms and conditions during the Promotion Exclusive Negotiation Period, then the Parties shall promptly thereafter enter into a definitive agreement reflecting such terms. If the Parties fail to reach agreement during the Promotion Exclusive Negotiation Period on terms and conditions pursuant to which Regeneron would receive a right to Promote such Product in the Kiniksa Field in the U.S., then, subject to Section 2.10.4 (Failure to Agree; Promotion Thirty Party Offers), Kiniksa shall thereafter be free to engage in U.S. Promotion Negotiations with Third Parties for, and enter into U.S. Third Party Promotion Agreements with, Third Parties with respect to the Promotion of such Product in the U.S. without further obligations under this Section 2.10 (Regeneron ROFN to Promote Products in the U.S.).

2.10.4 **Failure to Agree; Promotion Third Party Offers**. If Regeneron provides a Promotion ROFN Exercise Notice in good faith pursuant to Section 2.10 (Regeneron ROFN to Promote Products in the U.S.), but subsequently the Parties are unable to agree on the terms of a U.S. Third Party Promotion Agreement during the Promotion Exclusive Negotiation Period, then if Kiniksa receives a written offer from a

Third Party and is prepared to accept such offer ("Third Party Offer") within [***] after the expiration of the Promotion Exclusive Negotiation Period, then prior to accepting such Third Party Offer,

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Kiniksa shall deliver written notice to Regeneron informing Regeneron of the financial and material terms of the Third Party Offer. No later than [***] after receiving such Third Party Offer, Regeneron shall have the right to match the financial and material terms of such Third Party Offer if the terms of such Third Party Offer are as a whole substantially equivalent to, or less favorable to Kiniksa than the last terms offered by Regeneron, and in such case, the Parties shall execute a binding term sheet and shall use good faith efforts to negotiate a definitive agreement reflecting the foregoing arrangement as promptly as is practicable thereafter.

2.11 **Regeneron ROFN to Purchase Rights to a Product**. Prior to the date of receipt of U.S. Marketing Approval, Kiniksa shall not assign its rights or obligations under this Agreement to any Third Party without Regeneron's consent as set forth in Section 17.9.1 (Assignment to Third Parties by Kiniksa). In addition, after the date of receipt of U.S. Marketing Approval, Kiniksa shall not enter into any negotiations or enter into any agreement with a Third Party pursuant to which Kiniksa would be permitted to assign its rights to the Product as permitted pursuant to Section 17.9.1 (Assignment to Third Parties by Kiniksa), in each case, except in compliance with this Section 2.11 (Regeneron ROFN to Purchase Rights to a Product). For clarity, this Section 2.11 (Regeneron ROFN to Purchase Rights to a Product) shall not apply to the transfer of the Product by way of a Change of Control of Kiniksa which shall be covered by Section 2.12 (Change of Control of Kiniksa).

2.11.1 **Sale ROFN**. On a country-by-country basis with respect to the Territory Kiniksa hereby grants to Regeneron a right of first negotiation in the event Kiniksa wishes to enter an arrangement to assign its rights to a Product to Develop and Commercialize the Product in the Kiniksa Field in the Territory (each, a "Sale ROFN"). In such event, Kiniksa will deliver a notice to Regeneron of its desire to assign to such Product (the "Sale ROFN Notice") prior to Kiniksa engaging in any negotiations with, accepting any offer from, or entering into any agreement, with any Third Party to assign such rights (collectively, "Sale Negotiations").

2.11.2 **Sale ROFN Exercise Notice**. If the event Regeneron wishes to enter into exclusive negotiations with Kiniksa to obtain the rights that Kiniksa wishes to assign in the Kiniksa Field in the Territory, Regeneron shall provide Kiniksa with notice thereof ("Sale ROFN Exercise Notice") within [***] after Regeneron's receipt of the Sale ROFN Exercise Notice. If Regeneron fails to deliver to Kiniksa the Sale ROFN Exercise Notice within such [***] period, then Kiniksa shall thereafter be free to engage in Sale Negotiations with Third Parties for, and enter into a written agreement with respect to, the assignment of such rights to such Product (a "Sale Agreement") with any Third Party with respect to such Product without further obligations under this Section 2.11 (Regeneron ROFN to Purchase Rights).

2.11.3 **Sale Exclusive Negotiation Period**. In the event Regeneron delivers the Sale ROFN Exercise Notice with the [***] time period set forth in Section 2.11.2 (Sale ROFN Exercise Notice), the Parties will engage in good faith negotiations, and Kiniksa will permit Regeneron to conduct, and will permit Regeneron's conduct of, technical due diligence, for a period of [***] after delivery of the Sale ROFN Exercise Notice ("Sale Exclusive Negotiation Period") in an attempt to agree upon the terms and conditions pursuant to which Regeneron would receive rights to such Product in the Kiniksa Field in the Territory (or a portion thereof). If the Parties are able to reach agreement on such terms and conditions during the Sale Exclusive Negotiation Period, then the Parties shall promptly thereafter enter into a definitive agreement reflecting such terms. If the Parties fail to reach agreement during the Sale Exclusive Negotiation Period on terms and conditions for Regeneron's acquisition of rights in the Kiniksa Field in the Territory (or a portion thereof), then Kiniksa shall thereafter be free to engage in Sale Negotiations with Third Parties for, and enter into Sale Agreements with any Third Party with respect to assignment of rights to such Product that is the subject of the Sale ROFN Notice without further obligations under this Section 2.11 (Regeneron ROFN to Purchase Rights), subject to Section 2.11.4 (Failure to Agree; Sale Third Party Offers).

2.11.4 **Failure to Agree; Sale Third Party Offers**. If Regeneron provides a Sale ROFN Exercise Notice in good faith pursuant to Section 2.11.2 (Sale ROFN Exercise Notice), but subsequently the Parties are unable to agree on the terms of a Sales Agreement during the Sale Exclusive Negotiation Period, then if Kiniksa receives a Third Party Offer within [***] after expiration of the Sale Exclusive Negotiation Period, then prior to accepting such Third Party Offer, Kiniksa shall deliver written notice to Regeneron informing Regeneron of the financial and material terms of the Third Party Offer. Within [***] of receiving such Third Party Offer, Regeneron shall have the right to [***], and in such case, the Parties shall [***] reflecting the foregoing arrangement as promptly as is practicable thereafter. Kiniksa shall not grant any rights to Third Parties with respect to any Product that would conflict with Regeneron's rights pursuant to this Section 2.11.4 (Failure to Agree; Sale Third Party Offers).

2.12 **Change of Control of Kiniksa**. During the period commencing on the Effective Date and ending upon the earlier of (a) [***], or (b) [***], if Kiniksa's management approves a binding term sheet or approves the initiation of the negotiation of a definitive agreement, in each case, with a Third Party with respect to a Change of Control of Kiniksa, then Kiniksa shall immediately deliver notice to Regeneron stating that the foregoing has occurred.

ARTICLE 3
TRANSFERS AND REGENERON CONSULTATION ASSISTANCE; ASSUMED LIABILITIES

3.1 **Transfers on the Effective Date.**

3.1.1 **Product Records**. On or as reasonably practicable following the Effective Date, Regeneron will use Commercially Reasonable Efforts to provide Kiniksa or its designee with access to all Product Records in Regeneron's possession or that may be readily available to Regeneron, excluding Product Records relating to the Retained Field or the Regeneron Retained EEO Field. In addition, no later than [***] days after the Effective Date, Regeneron shall deliver or cause to be delivered to Kiniksa electronically or EXW (Incoterms 2010) (if in physical form) all Product Records in Regeneron's possession or that may be readily available to Regeneron, excluding Product Records relating to the Regeneron Field or the Regeneron Retained EEO Field.

3.1.2 **Existing Contracts in the Kiniksa Field**. As of the Effective Date, neither Regeneron nor any of its Affiliates is a party to any Third Party agreements (but excluding any agreements related to investigator initiated studies for the Product in the Territory that are covered by Section 3.5.2 (Existing Contracts in the Regeneron Field and all investigator initiated studies)) that are specific to and exclusively related to the Development or Commercialization of the Product in the Kiniksa Field in the Territory

3.1.3 **Regulatory and Safety Information**. On or as reasonably practicable following the Effective Date, Regeneron will use Commercially Reasonable Efforts to provide Kiniksa or its designee with access to (a) Regulatory Documentation and Approvals, in all cases, specifically and exclusively relating to the Development of the Product in the Regeneron Field, and (b) the global safety database for the Product and copies of all material safety data and other product data and information related to the Product in the Regeneron Field, in each of clauses (a) and (b), in Regeneron's possession and Control.

3.2 **Obligations Prior to Transfer of U.S. BLA**.

3.2.1 **Access to U.S. BLA; Cooperation**. No later than [***] months prior to the anticipated date of BLA Filing, or such earlier time as may be requested by Kiniksa and agreed to by Regeneron, such agreement not to be unreasonably, withheld, conditioned or delayed, for regulatory purposes in connection with meetings with, or submissions to, the FDA, in a location agreed to by Regeneron and reasonably acceptable to Kiniksa, Regeneron will make arrangements for, and permit, Kiniksa or its designee to access (a) all Product Manufacturing Records relating to the Kiniksa Field in Regeneron's possession or that may be readily available to Regeneron and (b) the U.S. BLA, including making available to Kiniksa or its designee all changes and amendments to such U.S.

BLA. In connection with such review by or on behalf of Kiniksa, Regeneron will use reasonable efforts to ensure that Regeneron personnel with relevant experience, including with respect to CMC and other Manufacturing activities related to the Product, are reasonably available to Kiniksa to answer questions and discuss such Product Manufacturing Records and the U.S. BLA, including any changes or amendments thereto since its original filing with the FDA.

3.2.2 **Meetings with FDA**. Without limiting the timing and conditions for the transfer of the U.S. BLA as set forth in Section 3.3 (Transfers in Connection with the U.S. BLA Transfer), prior to the U.S. BLA Transfer Date, upon Kiniksa's reasonable request, Regeneron will request meetings with the FDA to discuss and seek guidance regarding the U.S. BLA in its current form and all information and data proposed by Kiniksa to be included with its supplemental BLA in the Kiniksa Field. Regeneron will use reasonable efforts to ensure that Regeneron personnel with relevant experience, including with respect to CMC and other Manufacturing activities related to the Product, attend such meeting with the FDA, along with Kiniksa or its designees. Regeneron's Fully-Burdened Costs for providing services pursuant to this Section 3.2.2 (Meeting with FDA) shall be considered Other Shared Expenses.

3.2.3 **Authorization**. Regeneron shall file with the FDA or other Regulatory Authority any documents required, or take any other action necessary, to permit Kiniksa to Develop the Product in the Kiniksa Field in the Territory before the U.S. BLA Transfer Date.

3.3 **Transfers in Connection with the U.S. BLA Transfer.**

3.3.1 **Transfer of U.S. BLA**. As soon as practicable after a Kiniksa Field Positive Clinical Readout, the Parties shall file with the FDA all information required in order to transfer the U.S. BLA from Regeneron to Kiniksa, including any authorization letters, notices, and letters of acceptance required to transfer the U.S. BLA from Regeneron to Kiniksa. Regeneron shall file with the FDA the information required of a former owner of a BLA, and Kiniksa shall file with the FDA the information required of a new owner of a BLA, in each case, at each Party's own expense. Each of Regeneron and Kiniksa also agree to take any actions required by any Governmental Authority to effect the transfer of the U.S. BLA from Regeneron to Kiniksa, and hereby further agree to cooperate with each other in order to effectuate the

transfer of the U.S. BLA. Any fees imposed by Regulatory Authorities or Applicable Law related to the transfer or the recording thereof shall be at Kiniksa's expense. After transfer of the U.S. BLA from Regeneron to Kiniksa, but prior to the date of receipt of U.S. Marketing Approval, Kiniksa shall [***].

3.3.2 **Regulatory and Safety Information**. No later than [***] days after the U.S. BLA Transfer Date, and subject to Kiniksa's reasonable assistance, to the extent legally permissible (including to the extent permitted under Regeneron's

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obligations to Third Parties on the U.S. BLA Transfer Date), Regeneron shall (a) transfer and assign to Kiniksa or Kiniksa's designee Regeneron's rights, title, and interests in and to all Regulatory Documentation and Approvals [***] related to the Development of the Product in the Regeneron Field in the Territory, and (b) subject to an audit reasonably satisfactory to Regeneron as set forth in Section 7.6.2 (Pharmacovigilance Audit), transfer to Kiniksa or Kiniksa's designee the global safety database for the Product and copies of all such material safety data and other product data and information, in each case, in Regeneron's possession and Control. In the event of (i) failure to obtain assignment, or (ii) with respect to regulatory items that would otherwise fall within the foregoing clauses (i) or (ii), but for such materials not [***] related to a Product, but nonetheless that are necessary for the Development or Commercialization of such Product in the Kiniksa Field, in each case ((i) and (ii)), Regeneron hereby consents and grants to Kiniksa the right to reference any such item solely with respect to the Development or Commercialization of such Product in the Kiniksa Field.

3.4 **Obligations and Meetings with the FDA After Transfer of U.S**. BLA. After transfer of the U.S. BLA from Regeneron to Kiniksa in accordance with Section 3.3.1 (Transfer of the U.S. BLA):

3.4.1 Kiniksa shall file with the FDA or other Regulatory Authority any documents required, or take any other action necessary, to permit Regeneron to (a) Commercialize the Product in the Regeneron Field in the Territory before the date of receipt of U.S. Marketing Approval, or (b) Develop and Commercialize (including interaction with payors as appropriate) the Product in the Regeneron Retained EEO Field both before and after the date of receipt of U.S. Marketing Approval. Notwithstanding the foregoing, none of the foregoing required actions are intended to diminish and shall not limit Kiniksa's rights to Develop or Commercialize the Product in the Kiniksa Field;

3.4.2 Kiniksa shall not voluntarily initiate any communications with the FDA or respond to the FDA with respect to the Product (a) in the Regeneron Field before the date of receipt of U.S. Marketing Approval, or (b) in the Regeneron Retained EEO Field both before and after the date of receipt of U.S. Marketing Approval, without consultation and coordination with, [***] Regeneron, *unless* the U.S. BLA holder is required under Applicable Law or by the FDA to initiate such communications [***]; and

3.4.3 After transfer of the U.S. BLA from Regeneron to Kiniksa in accordance with Section 3.3.1 (Transfer of the U.S. BLA), upon Regeneron's request, Kiniksa shall request meetings with the FDA on Regeneron's behalf and Kiniksa shall attend such meetings upon Regeneron's request, to the extent Kiniksa's presence is necessary as the holder of the U.S. BLA and in such event, Kiniksa shall attend any such meetings as an observer, in each case, with respect to the

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Product (a) in the Regeneron Field before the date of receipt of U.S. Marketing Approval, or (b) in the Regeneron Retained EEO Field both before and after the date of receipt of U.S. Marketing Approval. Notwithstanding the foregoing, nothing in this Section 3.4 (Obligations and Meetings with the FDA After Transfer of the U.S. BLA) will prohibit Kiniksa from requesting or attending any meeting with the FDA related to the Regeneron Field or the Regeneron Retained EEO Field, to the extent such meeting is (a) required to be held with the holder of the U.S. BLA, or (b) related to the Kiniksa Field. The Parties will consult and cooperate with each other with respect to any such meeting with the FDA and attempt to reach all decisions related thereto by consensus, but Kiniksa will have final decision making authority if the Parties are unable to agree with respect to any matter related thereto (i) if the holder of the U.S. BLA is required under Applicable Law or by the FDA to make such decision, or (ii) related to the Kiniksa Field.

3.5 **Transfers Upon Receipt of U.S. Marketing Approval.**

3.5.1 **Product Records**. On or as reasonably practicable following the date of receipt of U.S. Marketing Approval, Regeneron will use Commercially Reasonable Efforts to provide Kiniksa or its designee with access to all Product Records relating to the Regeneron Field in Regeneron's possession or that may be readily available to Regeneron. In addition, no later than [***] days after the date of receipt of U.S. Marketing Approval, Regeneron shall deliver or cause to be delivered to Kiniksa EXW (Incoterms 2010) (if in physical form) all Product Records relating to the Regeneron Field in Regeneron's possession or that may be readily available to Regeneron.

3.5.2 **Existing Contracts**. All Third Party agreements to which Regeneron or its Affiliates is a party that are [***] related to the Development or Commercialization of the Product (including specialty pharmacy agreements) in the Regeneron Field in the Territory as of the Effective

Date and all agreements related to investigator initiated studies in the Territory whether in the Kiniksa Field or the Regeneron Field are set forth on Schedule 3.5.2 (the agreements set forth on Schedule 3.5.2, as updated in accordance with this Section 3.5.2 (Existing Contracts), the "Existing Contracts"). At least [***] days prior to the expected date of receipt of U.S. Marketing Approval, Regeneron will provide to Kiniksa an updated version of Schedule 3.5.2 (which for clarity will include any agreements that are [***] related to (a) investigator initiated studies in the Kiniksa Field or the Regeneron Field or (b) Regeneron Exploratory Clinical Studies in the Kiniksa Field, in each case, entered into by Regeneron pursuant to Section 3.6 (Investigator Initiated Studies and Regeneron Exploratory Clinical Studies).Within [***] days of receiving such updated Schedule 3.5.2 pursuant to the previous sentence, Kiniksa shall inform Regeneron in writing whether it wishes to be assigned any or all of such Existing Contracts set forth on Schedule 3.5.2 (as updated). On the date of receipt of U.S. Marketing Approval, at

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Kiniksa's option, to the extent requested by Kiniksa pursuant to the previous sentence and subject to any required consents, Regeneron will assign and transfer to Kiniksa any so requested Existing Contracts set forth on Schedule 3.5.2 (as updated). If any consent cannot be obtained with respect to any Existing Contract set forth on Schedule 3.5.2 (as updated), then Regeneron shall, and shall cause its Affiliates to, use reasonable efforts [***] to obtain for Kiniksa substantially all of the practical benefit and burden under such Existing Contracts and pass through such benefits to Kiniksa [***]. Regeneron will be solely responsible and liable for fulfillment of any commitments, and will pay any amounts due, pursuant to all agreements entered into by Regeneron related to the Product in the Regeneron Field prior to the date of receipt of U.S. Marketing Approval. For clarity, [***].

3.5.3 **Commercial Information**. On or as reasonably practicable following the date of receipt of U.S. Marketing Approval, Regeneron will use Commercially Reasonable Efforts to provide Kiniksa or its designee with access to all Commercial Information for Kiniksa to conduct Promotional activities of the Product in the Regeneron Field in the Territory, in each case in Regeneron's possession and Control. In addition, no later than [***] days after the date of receipt of U.S. Marketing Approval, Regeneron shall deliver or cause to be delivered to Kiniksa electronically or EXW (Incoterms 2010) (if in physical form) all such Commercial Information, in each case in Regeneron's possession and Control. Subject to Kiniksa's reasonable assistance, to the extent legally permissible, Regeneron shall (a) transfer and assign to Kiniksa or Kiniksa's designee Regeneron's rights, title, and interests in and to all Approvals, including Pricing Approvals [***] relating to the Commercialization of the Product in the Regeneron Field in the Territory, in each case, in Regeneron's possession and Control (to the extent not previously transferred to Kiniksa). Additionally, upon Kiniksa's written request delivered at any time within [***] months prior to the expected date of U.S. Marketing Approval, Regeneron shall provide Kiniksa with the then-current status of inventory of the Product held by its distributor for the Regeneron Field.

3.5.4 **Additional Safety Information**. No later than [***] days after the date of receipt of U.S. Marketing Approval, Regeneron shall, subject to Kiniksa's reasonable assistance, to the extent legally permissible [***], transfer to Kiniksa or Kiniksa's designee copies of all Product safety data in Regeneron's possession and Control (including all Product Records) that have not already been transferred to Kiniksa. In the event of any failure to obtain assignment, Regeneron hereby consents and grants to Kiniksa the right to reference any such item solely with respect to the Development, Commercialization, or the Manufacture of such Product in the Kiniksa Field and the Regeneron Field.

3.5.5 **Promotional Activities**. As of the Effective Date, Regeneron and its Affiliates are not performing any Promotional activities for the Product in the Regeneron

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Field. To the extent that after the Effective Date, but before the date of U.S. Marketing Approval, Regeneron performs Promotional activities in the Regeneron Field, then Regeneron shall conduct all such Promotional activities in the Regeneron Field in accordance with Applicable Law and shall not make any false or misleading statements about the Product or Promote the Product other than in accordance with the applicable Approvals and the approved labeling at such time. Upon Kiniksa's request, delivered no later than [***] days following the date of receipt of U.S. Marketing Approval, Regeneron shall transfer to Kiniksa all Promotional activities for the Product in the Territory in the Regeneron Field (as defined as of the Effective Date) at Kiniksa's direction, and the costs incurred with such transfer shall be considered Other Shared Expenses.

3.5.6 **Transfers in Connection with a Retained Field Event**. In connection with a Retained Field Event, Regeneron shall use reasonable efforts [***] to provide Kiniksa with the information, agreements, and assistance with respect to the Product as set forth in Section 3.1 (Transfers on the Effective Date), Section 3.5.2 (Existing Contracts), Section 3.5.3 (Commercial Information), and Section 3.5.5 (Promotional Activities), [***] as applicable, to the extent such Product Records are in Regeneron's possession or are readily available to Regeneron.

3.6 **Investigator Initiated Studies and Regeneron Exploratory Clinical Studies**. Prior to the date of receipt of U.S. Marketing Approval, Regeneron (through the JSC pursuant to Section 4.1.3(s)) shall (a) be responsible for coordinating any Product requests for investigator initiated studies in the U.S. in all Indications and (b) have the right to perform clinical studies for the Product in all Indications ("Regeneron Exploratory Clinical Studies") (without any right to Commercialize the Product in the Kiniksa Field); provided that Kiniksa shall have the right to approve any Product requests for

investigator initiated studies in the U.S. and Regeneron's requests to perform Regeneron Exploratory Clinical Studies, in each case, outside of the Regeneron Field and outside the Regeneron Retained EEO Field, such approval not to be unreasonably withheld, conditioned, or delayed. In addition, [***]. After the date of receipt of U.S. Marketing Approval, Kiniksa shall be responsible for coordinating any Product requests for investigator initiated studies in the Territory and shall have the right to approve any Product requests for investigator initiated studies in the Territory for testing outside of the Retained Field. After the date of receipt of U.S. Marketing Approval, Regeneron shall not have the right to perform Regeneron Exploratory Clinical Studies in the Kiniksa Field.

3.7 **Product Records Prohibited to be Provided**. To the extent that the transfer or delivery to Kiniksa of any Product Record is prohibited by any Applicable Law or would require any governmental or Third Party authorizations, consents, or waivers, and Regeneron has used reasonable efforts to obtain, but has not obtained, such authorizations, consents, or waivers prior to the date such Product Record is required to be transferred to Kiniksa pursuant to Section 3.1.1 (Transfers on the Effective Date; Product Records), or Section 3.5.1 (Transfers on the Date of Receipt of U.S. Marketing Approval; Product Records), as

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applicable, then the failure to transfer or deliver such Product Records shall not constitute a breach of this Agreement by Regeneron. In such event, Regeneron and Kiniksa shall reasonably cooperate with each other to promptly obtain such authorizations, consents, or waivers. Pending such authorization, consent, or waiver, Regeneron and Kiniksa shall reasonably cooperate with each other in any agreeable, reasonable, and lawful arrangements designed to provide to Kiniksa the benefits of use of such Product Records.

3.8 **Ongoing Updates**. Following each date on which Regeneron is required to assign to Kiniksa applicable Product Records and Commercial Information pursuant to this ARTICLE 3 (Transfers and Regeneron Consultation Assistance; Assumed Liabilities), as applicable, thereafter Regeneron will promptly share with Kiniksa any material additions to such Product Records and Commercial Information that come into its possession that have not been previously provided to Kiniksa pursuant to this ARTICLE 3 (Transfers and Regeneron Consultation Assistance; Assumed Liabilities). In the event Regeneron conducts Regeneron Exploratory Clinical Studies, Regeneron shall provide the JSC with quarterly updates regarding the status of such Regeneron Exploratory Clinical Studies, and upon completion of such Regeneron Exploratory Clinical Studies, promptly after such data and information are available, transfer and assign the Product Records and Regulatory Documentation arising out of such Regeneron Exploratory Clinical Studies to Kiniksa.

3.9 **Regeneron Development in the Regeneron Retained EEO Field**.

3.9.1 **Notice by Regeneron**. Regeneron shall inform Kiniksa in writing if Regeneron determines to Develop the Product in the Regeneron Retained EEO Field.

3.9.2 **Coordination**. In the event Regeneron Develops the Product in the Regeneron Retained EEO Field, the following provisions shall apply:

(a) Prior to initiating any clinical studies in the Regeneron Retained EEO Field (regardless of whether such study is a Regeneron Exploratory Clinical Study or an investigator initiated study), Regeneron will provide a summary of the applicable study protocol to Kiniksa and any material changes thereto. If such study is a Regeneron Exploratory Clinical Study, then Regeneron will provide to Kiniksa an initial, high-level summary at such time that Regeneron initiates development of a complete protocol for any such study.

(b) Both Parties will cooperate with each other to develop and follow specific procedures to be agreed upon to coordinate the exchange of necessary regulatory information regarding the Product Developed and Commercialized by or on behalf of Kiniksa in the Kiniksa Field in the Territory and from the Product Developed and Commercialized by or on behalf of Regeneron in the Regeneron Retained EEO Field. For the purpose of clarity, such regulatory information shall be limited to the extent that such regulatory information may reasonably be expected to

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have an impact on the Development or Commercialization of the Product Developed and Commercialized by or on behalf of Kiniksa in the Kiniksa Field in the Territory and the Product Developed and Commercialized by or on behalf of Regeneron in the Regeneron Retained EEO Field.

(c) Kiniksa agrees to promptly disclose to Regeneron all information related to the Product Developed and Commercialized by or on behalf of Kiniksa under this Agreement in the Kiniksa Field in the Territory to the extent that such information may reasonably be expected to have an impact on the Development or Commercialization of the Product in the Regeneron Retained EEO Field. Regeneron agrees to promptly disclose to Kiniksa all information related to the Product Developed and Commercialized by or on behalf of Regeneron under this Agreement in the Regeneron Retained EEO Field to the extent that such information may

reasonably be expected to have an impact on the Development or Commercialization of the Product in the Kiniksa Field in the Territory.

(d) Kiniksa shall notify Regeneron if any data regarding the Product in the Kiniksa Field generated under this Agreement is submitted to Regulatory Authorities or Governmental Authorities in support of the Product in the Territory to the extent that such data may reasonably be expected to have an adverse impact on the Product in the Regeneron Retained EEO Field. In the event that Kiniksa's regulatory strategy pertaining to such data may reasonably be expected to have an adverse impact on Regeneron's regulatory strategy used to support in the Product in the Regeneron Retained EEO Field, Regeneron and Kiniksa shall discuss and agree to the proposed regulatory strategy pertaining to such data in advance of Kiniksa's (or its Affiliates' or licensees') communication with Regulatory Authorities or Governmental Authorities regarding such data; provided that Kiniksa shall have the final decision making authority with respect to such regulatory strategy and any such communications. Regeneron shall notify Kiniksa if any data regarding the Product in the Regeneron Retained EEO Field generated under this Agreement is submitted to Regulatory Authorities or Governmental Authorities in support of the Product to the extent that such data may reasonably be expected to have an adverse impact on the Product in the Kiniksa Field in the Territory. In the event that Regeneron's regulatory strategy pertaining to such data may reasonably be expected to have an adverse impact on Kiniksa's regulatory strategy used to support in the Product in the Kiniksa Field in the Territory, Regeneron and Kiniksa shall discuss and agree to the proposed regulatory strategy pertaining to such data in advance of Regeneron's (or its Affiliates' or licensees') communication with Regulatory Authorities or Governmental Authorities regarding such data; provided that Regeneron shall have the final decision making

authority with respect to such regulatory strategy and any such communications, subject to Section 3.4.2 (Obligations and Meetings with the FDA After Transfer of U.S. BLA.

3.10 **Technical Consultation and Transition Services**.

3.10.1 **Provision of Services**. Following the Effective Date, with respect to the Kiniksa Field, and following the date of receipt of U.S. Marketing Approval with respect to the Regeneron Field, in each case, upon Kiniksa's written request, Regeneron shall provide Kiniksa or its designee with reasonable technical consultation to explain the Product Records and the Regeneron Know-How, and transition support services by Regeneron personnel with the relevant experience to the extent necessary to ensure a complete, orderly, and successful transition of Development activities related to the Product in the Kiniksa Field and uninterrupted Commercialization operations related to the Product in the Regeneron Field, and in case, to the extent performed by Regeneron in the Regeneron Field as of the date of U.S. Marketing Approval (the "Technical Consultation and Transition Services").

3.10.2 **Service Period**. Regeneron's obligation to provide Technical Consultation and Transition Services pursuant to Section 3.10.1 (Provision of Services) shall expire upon (a) upon the [***], and (b) upon the [***]. Additionally, [***].

3.10.3 **Service Costs and Expenses**. Regeneron shall provide to Kiniksa up to [***] of Technical Consultation and Transition Services without charge with respect to any FTE Costs, and Kiniksa shall be responsible for additional Technical Consultation and Transition Services in excess of [***] in connection therewith. The Fully-Burdened Cost to Regeneron for any such excess Technical Consultation and Transition Services shall be considered Other Shared Expenses. At Kiniksa's request, Regeneron shall provide a good faith estimate of its Fully-Burdened Cost for providing Technical Consultation and Transition Services.

3.11 **Assumed Liabilities and Obligations and Excluded Liabilities and Obligations**.

3.11.1 **Effective Date**. As of the Effective Date, Kiniksa hereby assumes and agrees to discharge promptly as they become due any and all liabilities and obligations related to or arising from (a) Development of the Product for Commercialization in the Kiniksa Field in the Territory (excluding for Regeneron Exploratory Clinical Studies and for investigator initiated studies enabled by Regeneron in the U.S. in accordance with Section 3.6 (Investigator Initiated Studies and Regeneron Exploratory Clinical Studies)), and (b) the Commercialization of the Product in the Kiniksa Field in the Territory, in each case ((a) and (b)), subject to Regeneron's obligation to Manufacture the Product as set forth under this Agreement and the Supply Agreements. Kiniksa shall not assume any liabilities and obligations related to or arising from the Development, Commercialization,

or Manufacture of the Product for any period ending on or prior to the Effective Date.

3.11.2 **U.S. BLA Transfer Date**.

(a) Prior to the U.S. BLA Transfer Date. For all periods ending on or prior to the U.S. BLA Transfer Date, Regeneron shall maintain and agrees to discharge promptly as they become due, any and all liabilities and obligations related to or arising from the maintenance of the U.S. BLA and Kiniksa shall not assume any liabilities and obligations related to the foregoing.

(b) After the U.S. BLA Transfer Date. For all periods commencing after the U.S. BLA Transfer Date, Kiniksa shall maintain and agrees to discharge promptly as they become due, any and all liabilities and obligations related to or arising from the maintenance of the U.S. BLA, including those obligations with respect to making filings associated with Product Changes in accordance with Section 8.8 (Product Changes), and Regeneron shall not assume any liabilities and obligations related to the forgoing, except for obligations to prepare filings associated with Product Changes in accordance with Section 8.8 (Product Changes).

3.11.3 **U.S. Marketing Approval**. For all periods after the date of receipt of U.S. Marketing Approval, Kiniksa hereby assumes and agrees to discharge promptly as they become due any and all liabilities and obligations related to or arising from (a) the Development of the Product for Commercialization in the Regeneron Field (as incorporated into the Kiniksa Field), and (b) the Commercialization of the Product in the Regeneron Field (as incorporated into the Kiniksa Field), in each case ((a) and (b)), subject to Regeneron's obligation to Manufacture the Product as set forth under this Agreement and the Supply Agreements. Kiniksa shall not assume any, and Regeneron shall retain all, liabilities and obligations related to or arising from (i) the Development of any Product for Commercialization in the Regeneron Field, (ii) the Commercialization of the Product in the Regeneron Field, or (iii) the Manufacture of any Product, in each case ((i) — (iii)), for any period ending on or prior to the date of receipt of U.S. Marketing Approval.

3.11.4 [***]. For all periods after the date of the [***], Kiniksa hereby assumes and agrees to discharge promptly as they become due any and all liabilities and obligations related to or arising from (a) the Development of the Product for Commercialization in the [***] (as incorporated into the Kiniksa Field) in the Territory, and (b) the Commercialization of the Product in the [***] (as incorporated into the Kiniksa Field) in the Territory, in each case ((a) and (b)), subject to Regeneron's obligation to Manufacture such Product as set forth under this Agreement and the Supply Agreements. Kiniksa shall not assume any, and Regeneron shall retain all, liabilities and obligations related to or

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arising from (i) the Development of any Product for Commercialization in the [***], (ii) the Commercialization of any Product in the [***], or (iii) the Manufacture of the Product, in each case ((i) — (iii)), for any period ending on or prior to the date of the [***].

3.11.5 **Regeneron Retained Liabilities**. Kiniksa shall not assume any, and Regeneron shall retain all, liabilities and obligations related to or arising from the performance of Regeneron Exploratory Clinical Studies, the Development of the Product for Commercialization in the Regeneron Retained EEO Field and Commercialization of the Product in the Regeneron Retained EEO Field.

3.11.6 **Manufacturing Technology Transfer Event**. After the date of completion of a Fill/Finish Technology Transfer (other than in connection with a Formulated Bulk Technology Transfer), to the extent performed by or behalf of Kiniksa (and for clarity, not by Regeneron under the Supply Agreements), Kiniksa will be responsible for any and all liabilities and obligations related to or arising from such fill/finish activities for the Product after such date. After the date of completion of any other Manufacturing Technology Transfer to the extent performed by and behalf of Kiniksa (and for clarity, not by Regeneron under the Supply Agreements), Kiniksa will be responsible for any and all liabilities and obligations related to or arising from the Manufacturing of the Product arising after such date.

ARTICLE 4
GOVERNANCE

4.1 **The Joint Steering Committee**.

4.1.1 **Formation, Composition, and Membership**. Promptly after the Effective Date, the Parties will establish a Joint Steering Committee (the "Joint Steering Committee" or "JSC"), which JSC shall consist of at least [***] senior representatives appointed by each of Regeneron and Kiniksa with experience appropriate for service on the JSC in light of the functions, responsibilities, and authority of the JSC and the status of Development or Commercialization of the Product being pursued hereunder from time to time. Each Party may replace its JSC members upon written notice to the other Party (which notice may be provided via email; *provided that* such replacement is a senior representative of such Party). The JSC will have two (2) co-chairpersons, one designated by each of Regeneron and Kiniksa. The roles and responsibilities of the JSC are set forth in this Agreement. From time to time, the JSC may establish working groups (each, a "Working Group") to oversee particular projects or activities, and each such Working Group shall be constituted and shall operate as the JSC for such matters. So long as Regeneron is Manufacturing any Product for Kiniksa under the Supply Agreement, a supply chain representative from each Party shall be represented on the JSC or a Working Group.

4.1.2 **Meetings of the JSC**. The JSC shall meet at least [***] until [***], and once every [***] thereafter during the duration of the Term, in each case, unless the JSC co-chairpersons otherwise agree. The first meeting of the JSC will be held at Regeneron's facilities in New York on a date agreed to by the Parties no later than thirty (30) days after the Effective Date. All JSC meetings may be conducted by telephone, video-conference, or in person as determined by the JSC co-chairpersons; *provided that* the JSC shall meet in person at least [***]. Unless otherwise agreed by the Parties, all in-person meetings of the JSC shall be held alternatively at Regeneron's facilities in New York and at Kiniksa's or it's Affiliate's offices in Massachusetts. Further, each co-chairperson shall be entitled to call meetings in addition to the regularly scheduled [***] meetings. The co-chairpersons, with the assistance of the Alliance Managers, shall coordinate activities to prepare and circulate an agenda in advance of each meeting and prepare and issue draft minutes from each meeting within [***] days thereafter and final minutes within [***] days thereafter. Upon the invitation of a Party, a reasonable number of other representatives of such Party may attend any JSC meeting as non-voting observers; *provided that* such additional representatives are under obligations of confidentiality and non-use applicable to the Confidential Information of the other Party that are at least as stringent as those set forth in ARTICLE 13 (Confidentiality). Each Party shall be responsible for all of its own personnel and travel costs and expenses relating to participation in JSC meetings.

4.1.3 **Duties of the JSC**. The JSC shall:

(a) review, discuss, and approve the group of countries that constitute the Region, as described in Section 1.173 (Region);

(b) subject to Section 4.1.3(e) (Duties of the JSC), review and discuss the overall Development strategy and Commercialization strategy of the Product in the Kiniksa Field in the Territory, and prior to the date of receipt of U.S. Marketing Approval, the overall Development strategy and Commercialization strategy of the Product in the Regeneron Field, including in relation to a Party's planned and actual Development and Commercialization activities in the Territory. It is the intention of the Parties that such discussions will serve as a means for Regeneron to provide input with respect to the Development and Commercialization of the Product in the Kiniksa Field in the Territory and for Kiniksa to provide input with respect to the Development and Commercialization of the Product in the Regeneron Field;

(c) subject to Section 4.1.3(e) (Duties of the JSC), review and discuss the Development and Commercialization of the Product being conducted under this Agreement and for general liaison and communication with

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respect to the activities being conducted under the Neovii License Agreement, subject to any confidentiality restrictions thereunder;

(d) review and discuss the costs required to support the Manufacture of the Product [***];

(e) review, discuss, and approve the Development Plan and Commercial Plan and each amendment thereto, as described in Section 5.4 (Development Plan) and Section 6.5 (Commercial Plan);

(f) prior to the [***], review, discuss, and approve any amendment to any Development Plan that includes (i) the commencement of any Development of a Product [***], (ii) any clinical trial for a Product that was not [***], or (iii) any clinical Development for a Product not [***]; *provided that* the JSC will not have the right to approve the budget associated with any of the matters set forth in the foregoing clauses (i), (ii), and (iii) included in the Development Plan or any update or amendment thereto, and Kiniksa will retain final decision making authority with respect to all such budgets, as described in Section 5.4 (Development Plan);

(g) review and discuss, and prior to [***] approve, (which for clarity may be done through an appropriate Working Group), protocols prepared by Kiniksa for clinical trials for any Product in the Kiniksa Field in the Territory;

(h) review and approve the initial Commercial Overhead Charge for each country or Region, and, no later than September 30 of each Contract Year, review and approve the updated Commercial Overhead Charge for each country or Region, as described in Section 6.7 (Commercial Overhead Charge) for the upcoming Contract Year;

(i) review and approve the initial Field Force FTE Rate for each country or Region, and, no later than September 30 of each Contract Year, review and approve the updated Field Force FTE Rate for each country or Region for the upcoming Contract Year, as described in Section 6.8 (Field Force FTE Rates);

(j) review and approve the initial FTE Rate (other than the Field Force FTE Rate) for each country or Region, and, no later than September 30 of each Contract Year, review and approve the updated FTE Rate for each country or Region for the upcoming Contract Year, as described in Section 6.9 (FTE Rates);

(k) establish procedures for the timely exchange of regulatory information, as described in Section 7.2 (Exchange of Regulatory Information);

(l) discuss Manufacturing and supply matters, including sharing information regarding Regeneron's expected Manufacturing capacity, Kiniksa's projected Product supply requirements, and the Manufacturing risk mitigation plan, as described in Section 8.9 (Risk Mitigation);

(m) review and discuss any Anticipated Supply Shortage, and determine whether to approve the engagement of any Third Party supply sources (and the identity thereof), as described in Section 8.13 (Notification and Discussion of Supply Issues);

(n) review and approve the form, format, and level of detail to be provided by the Parties in the reports to be provided under Section 9.4.1 (Periodic Reports);

(o) review and discuss the Projected Net Sales for the following Contract Year to be provided by Kiniksa, as described in Section 9.5.2(a) (Certain Shared Commercial Expenses [***];

(p) review and discuss each Subsequent Indication [***];

(q) review, discuss, and approve each Subsequent Indication [***];

(r) resolve any disputes with respect to the calculation of any Regeneron Profit Split Payment or Regeneron Third Party Proceeds Split Payment, or the contents of a Profit Payment Report or Third Party Proceeds Payment Report, as described in Section 9.9 (Resolution of Payment Disputes);

(s) prior to the date of U.S. Marketing Approval, review and discuss Product requests for investigator initiated studies in the U.S. for testing outside of the Regeneron Field and Regeneron requests to perform Regeneron Exploratory Clinical Studies in the Kiniksa Field, as described in Section 3.6 (Investigator Initiated Studies and Regeneron Exploratory Clinical Studies), and review and discuss the progress under and results of any investigator initiated studies related to any Product in the Regeneron Field in the Territory;

(t) review, discuss, and approve the entry into proposed Third Party Licenses (other than the Existing Licenses) by either Party, including those that require Third Party License Payments; *provided that* the JSC shall not withhold such approval if it determines that such Third Party License [***], as described in Section 2.8.3 (Freedom to Obtain New Third Party Licenses);

(u) resolve any disputes between the Parties with respect to the apportionment of Third Party License Payments between the Product in

the Kiniksa Field in the Territory and other countries, fields, products, or activities (as applicable, as described in Section 1.189(i) (Shared Commercial Expenses));

(v) discuss the use of the Existing Product Trademarks and the selection of New Product Trademarks, as described in Section 10.13.1 (Kiniksa's Use of Product Trademarks);

(w) resolve any disputes with respect to the results of any audit conducted under Section 11.2 (Audits and Adjustments), as described in Section 11.2.4 (Disputes);

(x) discuss potential Development activities related to delivering any Product in any alternative formulations, including a pre-filled syringe ("Formulation Development Activities"); and

(y) make any such decisions as are expressly allocated to the JSC under this Agreement or otherwise agreed by the Parties.

4.1.4 **Decision Making**. For those matters set forth in Section 4.1.3 (Duties of the JSC) that the JSC is only to review and discuss (and not approve), the JSC will review and discuss such matters at the next JSC (or appropriate Working Group meeting, if applicable), [***] the JSC (or appropriate Working Group, if applicable) will serve as a forum for communication, input, and discussion, but will not prevent a Party from taking action if such Party has final decision making authority and operational control with respect to such matters and such matter was brought before the JSC in accordance with Section 4.2.1 (Resolution of JSC Matters), but such matter will not be escalated pursuant to Section 4.2.2(b) (Escalation). For clarity, Kiniksa shall have decision making authority and operational control with respect to all matters pertaining to the Development or Commercialization of the Product in the Kiniksa Field in the Territory, including the implementation of the Development Plan or Commercial Plan without escalation pursuant to Section 4.2.2(b) (Escalation), subject to certain matters over which the JSC has an approval right, which matters shall be governed by Section 4.2.2(b) (Escalation) in the event that the JSC is unable to agree thereon. For those matters set forth in Section 4.1.3 (Duties of the JSC) that the JSC is to determine whether to approve, the JSC shall operate by consensus and for such matters, the representatives of each Party shall have collectively one (1) vote on behalf of such Party; *provided that* no such vote taken at a meeting shall be valid unless a representative of each Party is

present and participates in the vote. Notwithstanding the foregoing, each Party, in its sole discretion, by written notice to the other Party (which notice may be provided via email), may choose not to have representatives on the JSC, and in such case, the JSC to representatives of the other Party will make decisions on behalf of the JSC. The JSC must exercise its decision-making authority in good faith and in a commercially reasonable manner for the purpose of optimizing the

commercial potential of, and financial returns from, the Product in the Kiniksa Field in the Territory consistent with Commercially Reasonable Efforts. The Parties acknowledge and agree that the JSC will not have the power to amend any of the terms of this Agreement or take any action to expand or narrow the responsibilities of the JSC, in each case, other than by agreement of the Parties as set forth in Section 17.5 (Amendments).

4.2 **Resolution of JSC Matters**.

4.2.1 **Generally**. The Parties shall cause their respective representatives on the JSC to resolve all matters presented to them as expeditiously as possible and in a manner consistent with Commercially Reasonable Efforts, including all matters requiring approval.

4.2.2 **Unresolved Matters**.

(a) JSC Discussion. The JSC shall review and discuss the matters before it in good faith such that the perspectives of each Party's representatives on the JSC are given due consideration. For clarity, for matters not requiring JSC approval and on which the JSC is unable to reach agreement, the Party responsible for such matter under the Agreement may proceed if the JSC is unable to resolve such issue after a period: of (i) [***] Business Days after the JSC's discussion thereof in the event of an urgent matter, and (ii) [***] Business Days after the JSC's discussion thereof in the event of any other matter.

(b) Escalation. For those matters set forth in Section 4.1.3 (Duties of the JSC) [***], if the JSC is unable to make a decision due to a deadlock between the Regeneron vote on the one hand, and the Kiniksa vote on the other hand, after a period of [***] Business Days after the JSC's discussion thereof (or [***] Business Days after the JSC's discussion thereof in the event of an urgent matter) from the date on which such matter is submitted in writing to it for resolution pursuant to Section 4.2 (Resolution of JSC Matters), (any such matter, an "Unresolved Matter"), then either Party may require that the matter be submitted to the Executive Officers for a joint decision by submitting a written notice to the other Party formally requesting that the Unresolved Matter be resolved by the Executive Officers, and specifying the nature of the Unresolved Matter with sufficient specificity to permit adequate consideration by such Executive Officers. The Executive Officers shall diligently and in good faith work together and attempt to resolve the referred Unresolved Matter within [***] Business Days after receiving such written notification, or such longer period of time as the Executive Officers may agree in writing. All such referred Unresolved Matters shall require agreement of each Party's Executive Officer, and if such Executive Officers cannot resolve such Unresolved Matter within such

[***] Business Day period, or other period to which they may agree, then:

(i) *Matters Reserved for Decision by Regeneron*. Regeneron's Executive Officer shall have final decision-making authority with respect to any Unresolved Matter pertaining to (A) [***], or (B) during the period that Regeneron is Manufacturing the Product under the Supply Agreements, subject to the terms of the Supply Agreements, operational matters regarding the Manufacturing any Product, including the matters set forth in Section 4.1.3(m) (Duties of the JSC);

(ii) *Matters Reserved for Decision by Kiniksa*. Kiniksa's Executive Officer shall have final decision-making authority with respect to any Unresolved Matter pertaining to the Development or Commercialization of the Product in the Kiniksa Field in the Territory, including decision making for approval of, or operational matters regarding the implementation of, the Development Plan or the Commercial Plan or any update or amendment thereto, [***]. Notwithstanding the foregoing, [***];

(iii) *Determination of the Subsequent Indication* [***] in accordance with Section 9.5.3(b) (Expert Determination) and Section 17.1.2 (Financial Disputes); and

(iv) *Matters Subject to Dispute Resolution*. Section 17.1 (Disputes) shall apply with respect to the resolution of any other Unresolved Matter not covered by Section 4.2.2(b)(i) (Matters Reserved for Decision by Regeneron), Section 4.2.2(b)(ii) (Matters Reserved for Decision by Kiniksa), Section 4.2.2(b)(iii) (Determination of Subsequent Indication [***]) or Section 4.1.3(n). For clarity, neither Party shall have final decision making authority over Unresolved Matters regarding or affecting financial calculations hereunder, the form, format, and level of detail to be provided by the Parties in the reports to be provided under Section 9.4.1 (Periodic Reports), setting FTE Rates, setting the Commercial Overhead

Charge, without limiting Section 2.8.3 (Freedom to Obtain Third Party Licenses) the entry into any Third Party Licenses requiring Third Party License Payments (which licenses will be Third Party Licenses for purposes of this Agreement if required for freedom to operate), or regarding Legal Disputes or Financial Disputes, and in each case, such matters shall be resolved in accordance with Section 17.1 (Disputes).

4.3 **Alliance Management**. Each of Kiniksa and Regeneron shall appoint a senior representative who possesses a general understanding of research, clinical, regulatory,

manufacturing and marketing issues to act as its alliance manager, and each Party may replace such person upon notice (which notice may be via email) to the other Party ("Alliance Manager" or "Alliance Managers"). Each Alliance Manager shall be charged with creating and maintaining a collaborative work environment between the Parties. Each Alliance Manager will also be responsible for acting as a single-point of communication for seeking consensus both internally within the respective Party's organization and with the other Party's organization, including facilitating review of external corporate communications. The Alliance Managers shall continue to serve in their role during the Term of the Agreement.

ARTICLE 5
DEVELOPMENT OF PRODUCTS

5.1 **Development Responsibilities**. Subject to the terms of this Agreement, Kiniksa shall undertake Development activities with respect to the Product.

5.2 **Costs**. Kiniksa shall conduct Development of the Product under this Agreement at its sole cost and expense. For clarity, [***].

5.3 **Development Diligence Obligations**. Kiniksa shall use Commercially Reasonable Efforts to Develop the Product in the Kiniksa Field in the Territory. Without limiting the foregoing, Kiniksa shall use Commercially Reasonable Efforts to undertake and perform the Development activities set forth in the then-current Development Plan for the Product in the Kiniksa Field in a timely manner. Kiniksa shall conduct all such activities in compliance with Applicable Laws.

5.4 **Development Plan**. The initial Development Plan for the Product is set forth on Schedule 5.4 (the "Initial Development Plan"). Commencing in the [***] Contract Year, on an annual basis no later than September 30 of the then current Contract Year, or more frequently as may be determined by Kiniksa, Kiniksa shall prepare an updated Development Plan for the Product and provide such updated plan to the JSC for its review, discussion and approval pursuant to Section 4.1.3(e) (Duties of the JSC). Kiniksa shall consider in good faith comments by Regeneron regarding each update or amendment to the Development Plan; provided that Kiniksa shall have final discretion and decision making authority over the Development Plan and all updates or amendments thereto pursuant to Section 4.2.2(b)(ii) (Matters Reserved for Decision by Kiniksa). Notwithstanding the foregoing, Regeneron, and not Kiniksa, shall have final decision making authority over the matters [***], which matters, for clarity, will not include the budgets associated with any such matters and Kiniksa will retain final decision making authority regarding the budgets included in the Development Plan and each update or amendment thereto.

5.5 **Development Reports**. So long as Kiniksa is pursuing activities under the Development Plan or has future plans to conduct Development activities for the Product in the Kiniksa Field in the Territory, Kiniksa shall provide Regeneron with bi-annual written reports summarizing the status of all Development activities for the Product in the Kiniksa Field

in the Territory. Until the date of receipt of U.S. Marketing Approval, Regeneron shall provide Kiniksa with bi-annual written reports summarizing the status of all Development activities for the Product in the Regeneron Field.

5.6 **Regeneron Performance of Sample Bioanalysis**. At Kiniksa's written request, Regeneron shall perform sample bioanalysis of the Product in connection with clinical studies performed by Kiniksa for the Product in the Kiniksa Field in the Territory in accordance with the Development Plan using [***]: (a) [***] and (b) [***]. At Kiniksa's written request, in connection with the performance of a Phase III Trial for the Product in the Kiniksa Field in the Territory in accordance with the Development Plan, the Parties will discuss in good faith any assay optimization or assay validation activities to be performed by Regeneron. Regeneron's Fully-Burdened Costs for providing services pursuant to this Section 5.6 (Regeneron Performance of Sample Bioanalysis) shall be considered Other Shared Expenses.

5.7 **Regeneron Late Stage Development in the Regeneron Retained EEO Field**. In addition to any notice provided by Regeneron pursuant to Section 3.9.1 (Coordination), Regeneron shall inform Kiniksa in writing if Regeneron determines to conduct a Phase III Trial (or an adaptive clinical study that could convert into a Phase III Trial) of the Product in the Regeneron Retained EEO Field. Within [***] of Kiniksa's receipt of a notice from Regeneron pursuant to this Section 5.7 (Regeneron Late Stage Development in the Regeneron Retained EEO Field), the Parties shall negotiate in good faith and enter into an amendment to this Agreement to allow Regeneron to independently Develop and Commercialize the Product in the Regeneron Retained EEO Field in the Territory and, at the same time, allow Kiniksa to independently Develop and Commercialize the Product in the

Kiniksa Field in the Territory. In addition, in connection with such amendment, the Parties will evaluate whether certain [***] and will, if applicable, amend the Agreement to give effect to such [***]; *provided that* [***]. Lastly, such amendment may provide for additional coordination with respect to enforcement of Intellectual Property Rights and will provide for the rights of each Party if there is a dispute as to whether to initiate a recall or withdrawal of the Product after U.S. Marketing Approval. Notwithstanding the foregoing, (i) such amendment shall not diminish or limit any rights granted to Kiniksa under this Agreement, including Kiniksa's rights to determine pricing, reimbursement, and obtain Pricing Approvals in the Kiniksa Field and to otherwise Develop and Commercialize the Product in the Kiniksa Field and (ii) such amendment shall not diminish or limit any rights retained by Regeneron under this Agreement, including Regeneron's rights to determine pricing, reimbursement, and obtain Pricing Approvals in the Regeneron Retained EEO Field and to otherwise Develop and Commercialize the Product in the Regeneron Retained EEO Field.

ARTICLE 6
COMMERCIALIZATION OF THE PRODUCT

6.1 **Commercialization Responsibilities**. Subject to the terms of this Agreement, Kiniksa shall undertake Commercialization activities with respect to the Product.

6.2 **Sales and Distribution**. Until the date of receipt of U.S. Marketing Approval, for the Product, Regeneron shall be responsible for handling and collection of receivables, recording and booking sales, and handling returns, in each case, in each country in the Regeneron Field. Prior to the date of receipt of U.S. Marketing Approval, if Kiniksa receives any orders to purchase commercial Product in the Regeneron Field, then it shall refer such orders to Regeneron. After the date of receipt of U.S. Marketing Approval of the Product, Kiniksa shall be responsible for handling and collection of receivables, recording and booking sales, and handling returns, in each case, in each country in the Kiniksa Field in the Territory for Products sold or distributed after the date of receipt of U.S. Marketing Approval. After the date of receipt of U.S. Marketing Approval, if a Party receives any orders for any Product in the other Party's field or territory, then it shall refer such orders to the other Party.

6.3 **Costs**. Kiniksa shall conduct Commercialization of the Product under this Agreement at Kiniksa's sole cost and expense; provided that Kiniksa shall be entitled to deduct applicable costs and expenses in accordance with the Profit Split Arrangement as set forth in Section 9.4 (Sharing of Profits).

6.4 **Commercialization Diligence Obligations**. Kiniksa shall use Commercially Reasonable Efforts to undertake and perform the Commercialization activities set forth in the then-current Commercial Plan for the Product in the Kiniksa Field in the Territory in a timely manner. Kiniksa shall conduct all such activities in compliance with Applicable Laws. Without limiting the foregoing, after receipt of Marketing Approval for a Product in the Kiniksa Field in a country in the Territory, Kiniksa shall use Commercially Reasonable Efforts to Commercialize such Product in the Kiniksa Field in such country, including using Commercially Reasonable Efforts to obtain Pricing Approval (where applicable).

6.5 **Commercial Plan**. On a Major Market Country-by-Major Market Country basis, Kiniksa shall prepare a Commercial Plan for such Product, and, with sufficient time to meet the requirements set forth in the next sentence, provide such plan to the JSC for its review, discussion, and approval pursuant to Section 4.1.3(e) (Duties of the JSC). The JSC shall use good faith efforts to approve the Commercial Plan at least [***] prior to the Anticipated First Commercial Sale for a Product in such Major Market Country. Kiniksa shall consider in good faith comments by Regeneron regarding the Commercial Plan for such Product; provided that Kiniksa shall have final discretion and decision making authority over the Commercial Plan pursuant to Section 4.2.2(b)(ii) (Matters Reserved for Decision by Kiniksa). Thereafter, on an annual basis no later than September 30 of the then-current Contract Year, or more frequently as may be determined by Kiniksa, Kiniksa shall prepare an updated Commercial Plan for the Product for such Major Market

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Country and provide such updated plan to the JSC for its review, discussion, and approval pursuant to Section 4.1.3(e) (Duties of the JSC).

6.6 **Commercialization Report**. Kiniksa shall provide Regeneron with bi-annual written reports summarizing the status of all Commercialization activities for the Product in the Kiniksa Field in the Territory. Until the date of receipt of U.S. Marketing Approval, upon Kiniksa's written request, Regeneron shall provide Kiniksa with bi-annual written reports setting forth, for the Product in the Regeneron Field, sales for the prior six (6) month period covered by such report.

6.7 **Commercial Overhead Charge**. Kiniksa shall develop, and, with sufficient time to meet the requirements set forth in the next sentence, submit to the JSC for its review, discussion, and approval pursuant to Section 4.1.3(h) (Duties of the JSC) an initial Commercial Overhead Charge for each country or Region, which Commercial Overhead Charge will be applied on a quarterly basis in connection with the preparation of the Profit Payment Report in accordance with Section 9.4.1 (Periodic Reports). The JSC shall approve the Commercial Overhead Charge at least [***] prior to the anticipated Marketing Approval in such country or Region. For each upcoming Contract Year during the Term, Kiniksa shall develop an annual update to the Commercial Overhead Charge for each country or Region, and submit such updated amount to the JSC for its review and approval no later than September 30 of the then-current Contract Year pursuant to Section 4.1.3(h) (Duties of the JSC).

6.8 **Field Force FTE Rates**. Kiniksa shall develop, and with sufficient time to meet the requirements set forth in the next sentence, and submit to the JSC for its review, discussion, and approval pursuant to Section 4.1.3(i) (Duties of the JSC) an initial Field Force FTE Rate for such country or Region. The JSC shall approve the initial Field Force FTE Rate at least [***] prior to the anticipated Marketing Approval in such country or Region. Without limiting Section 9.6 (Adjustments to FTE Rate), for each upcoming Contract Year during the Term, Kiniksa shall develop an annual update to the Field Force FTE Rate for each country or Region, which rate will be adjusted by the applicable CPI Adjustment, and will submit such updated rate to the JSC for its review and approval no later than September 30 of the then-current Contract Year pursuant to Section 4.1.3(i) (Duties of the JSC) for the next Contract Year.

6.9 **FTE Rates**. In the first Contract Year that a Party is expected to incur FTE Costs in any country or Region in connection with performing activities related to any Product under this Agreement, such incurring Party shall develop and submit to the JSC for its review, discussion, and approval pursuant to Section 4.1.3(j) (Duties of the JSC) initial FTE Rates (other than a Field Force FTE Rate) for such country or Region. Without limiting Section 9.6 (Adjustments to FTE Rate), for each upcoming Contract Year during the Term, such Party shall develop an annual update to such FTE Rates (other than the Field Force FTE Rate) for each country or Region, which rate will be adjusted by the applicable CPI Adjustment, and will submit such updated rate to the JSC for its review

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and approval no later than September 30 of the then-current Contract Year pursuant to Section 4.1.3(j) (Duties of the JSC).

6.10 **Regeneron Acknowledgment**. Subject to Applicable Law in all cases, unless otherwise determined by Regeneron, in its sole discretion as communicated in writing to Kiniksa, all materials containing the Product Trademark shall state that (a) the Product was discovered by Regeneron and is part of a collaboration between Regeneron and Kiniksa, and (b) so long as Regeneron is Manufacturing Product under this Agreement, that the Product was manufactured by Regeneron. Kiniksa shall use reasonable efforts to file any documentation or obtain any consents required by Applicable Law in order to effectuate the previous sentence.

6.11 **Ex-Territory Sales; Export Monitoring**. Kiniksa and its Affiliates will use reasonable efforts to monitor and prevent exports of Products from in the Territory for Commercialization outside the Territory, and will monitor and prevent off-label use outside the Kiniksa Field (but otherwise in the Territory), in each case, using methods commonly used in the industry for such purpose. Kiniksa shall promptly inform Regeneron of any such exports of Products for Commercialization outside the Territory or off-label use outside the Kiniksa Field (but otherwise in the Territory), and the actions taken to prevent such exports or off-label use. Kiniksa agrees to take reasonable actions requested in writing by Regeneron that are consistent with Applicable Laws to prevent export of Products from its Territory for Commercialization outside the Territory or for off-label use outside the Kiniksa Field (but otherwise in the Territory). In connection with the Retained Field Event, Regeneron shall use reasonable efforts to obtain reciprocal assurances from Neovii with respect to the monitoring of exportation and off-label use by Neovii in the Kiniksa Field or in the Territory.

ARTICLE 7
CLINICAL AND REGULATORY AFFAIRS FOR PRODUCTS

7.1 **Ownership of Approval and Registration Filings**. Kiniksa will be solely responsible at its own cost and expense for preparing, applying for, coordinating, obtaining, and maintaining all Approvals and Registration Filings with respect to the Product for each Indication in the Kiniksa Field in the Territory where and when applicable, except as set forth in Section 8.8 (Product Changes). Kiniksa shall apply for and, when obtained, hold all Approvals and Registration Filings in its own name or in the name of one of its Affiliates. Kiniksa shall not assign or otherwise transfer any Approval or Registration Filing, or the right to apply therefore, to any Third Party without obtaining Regeneron's prior written consent, not to be unreasonably withheld, conditioned, or delayed.

7.2 **Exchange of Regulatory Information**. The Parties will establish procedures, through the JSC pursuant to Section 4.1.3(k) (Duties of the JSC), to ensure that the Parties exchange on a timely basis all necessary information to enable (a) each Party to comply with its regulatory obligations in connection with the Development, Manufacture, or Commercialization of the Product, including sharing relevant safety data, and filing updates or supplements with Regulatory Authorities, pharmacovigilance filings,

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Manufacturing supplements, and investigator notifications to Regulatory Authorities; and (b) each Party to comply with Applicable Laws in connection with the Development, Manufacture, or Commercialization of a Product. Each Party will provide prompt written notice to the other Party of any Approval of the Product for any Indication anywhere in the world.

7.3 **Certain Regulatory Materials**. (a) Prior to the date of receipt of U.S. Marketing Approval, and (b) at all times to the extent related to Manufacturing of the Product, if Regeneron is supplying Product to Kiniksa under the Supply Agreements, in each case ((a) and (b)), Kiniksa will provide Regeneron as promptly as practicable with written notice and copies of any material (i) draft filings; (ii) submissions to; and (iii) correspondence (including Approvals) with or to Regulatory Authorities that directly pertain to the Development or Commercialization of any Product in the Kiniksa Field in the Territory ("Regulatory Materials"). Kiniksa will use good faith efforts to afford Regeneron's representatives with

sufficient time to review and provide comments on such Regulatory Materials in advance of Kiniksa's submission thereof to Regulatory Authorities, and Kiniksa shall consider in good faith comments provided by Regeneron within [***] days of receipt by Regeneron, or such shorter period as may be required to satisfy requirements under Applicable Law. (A) Prior to the date of receipt of U.S. Marketing Approval, and (B) at all times to the extent related to Manufacturing of the Product, if Regeneron is supplying Product to Kiniksa under the Supply Agreements, in each case ((A) and (B)), Kiniksa will also, to the extent practicable and at Regeneron's expense, permit up to two (2) Regeneron representatives to attend (but not participate in) all material, pre-scheduled meetings, telephone conferences, or discussions with any Regulatory Authorities in the Territory at Regeneron's sole cost and expense; provided that such attendance is permitted by the applicable Regulatory Authority, but Kiniksa shall be under no obligation to provide translation services. Upon Regeneron's request, Kiniksa shall provide to Regeneron copies of all Registration Filings and Approvals (including supplements thereto) whether before or after the date of receipt of U.S. Marketing Approval. Without limiting the foregoing, Kiniksa will provide Regeneron with all material information, data, and materials reasonably necessary for Regeneron to participate in the review of the material filings and submissions referred to in this Section 7.3 (Certain Regulatory Materials) on a timely basis. For clarity, nothing in this Section 7.3 (Certain Regulatory Materials) shall limit the JSC's right to approve matters set forth in Section 4.1.3(f) (Duties of the JSC).

7.4 **Regulatory Events**. Each Party shall keep the other Party informed, as soon as possible, but no later than [***] after notification (or other time period specified in the Pharmacovigilance Agreement), of any action by, or notification or other information that it receives (directly or indirectly) from, any Regulatory Authority, Third Party, or other Governmental Authority that:

7.4.1 raises any material concerns regarding the safety or efficacy of any Product;

7.4.2 indicates or suggests a potential investigation or formal inquiry by any Regulatory Authority in connection with the Development, Manufacture, or Commercialization of any Product; *provided*, *however*, that each Party shall inform the other Party of the foregoing as soon as possible but in no event later than [***] after such Party's receipt of a notification referred to in this Section 7.4.2 (Regulatory Events); or

7.4.3 is reasonably likely to lead to a recall or market withdrawal of any Product anywhere in the Territory.

7.5 **Medical and Consumer Inquiries**. Prior to the date of receipt of U.S. Marketing Approval, (a) Regeneron shall be responsible for responding to medical questions or inquiries from members of the medical and paramedical professions and consumers regarding the Product in the Regeneron Field in the U.S. and Japan, and (b) Kiniksa shall be responsible for responding to medical questions or inquiries from members of the medical and paramedical professions and consumers regarding the Product in the Kiniksa Field in the Territory (except as otherwise mutually agreed to by the Parties in connection with Regeneron Exploratory Clinical Studies), in each case ((a) and (b)), in accordance with Applicable Law. After the date of receipt of U.S. Marketing Approval, Kiniksa shall be responsible for responding to medical questions or inquiries from members of the medical and paramedical professions and consumers regarding any Product in the Kiniksa Field in the Territory. If Kiniksa receives any such questions or inquiries about any Product in the U.S. and Japan prior to the date of receipt of U.S. Marketing Approval with respect to the Regeneron Field, then it will refer such questions or inquiries to Regeneron, and Regeneron will be responsible for responding thereto. If Regeneron receives any such questions or inquiries about any Product in the U.S. and Japan prior to the date of receipt of U.S. Marketing Approval in the Kiniksa Field or after the date of receipt of U.S. Marketing Approval in the Territory, then, in each case, it will refer such questions to Kiniksa or its designees, and Kiniksa will be responsible for responding thereto. At all times, Regeneron or its designee shall be responsible for responding to medical questions or inquiries from members of the medical and paramedical professions and consumers regarding any Product in the Retained Field. If, at any time, Kiniksa receives any such questions or inquiries about any Product in the Retained Field, then it will refer such questions to Regeneron or its designee, and Regeneron or its designee will be responsible for responding thereto. Nothing in this Section 7.5 (Medical and Consumer Inquiries) shall be construed to supersede the Pharmacovigilance Agreement and in the event of a conflict between this Section 7.5 (Medical and Consumer Inquiries) and the Pharmacovigilance Agreement, the Pharmacovigilance Agreement shall control.

7.6 **Pharmacovigilance**.

7.6.1 **Pharmacovigilance Agreement and Global Safety Database**. As soon as reasonably practicable after the Effective Date, but in any event within [***] after the Effective Date, or such later time as may be agreed by the Parties, but in any event prior to the commencement of any clinical activities by Kiniksa in

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the Territory, the Parties will develop and agree in writing upon a pharmacovigilance agreement ("Pharmacovigilance Agreement") that will include safety data exchange procedures governing the coordination of collection, investigation, reporting, and exchange of information concerning any adverse experiences and any product quality and product complaints involving adverse experiences, in each case, related to the Product, sufficient to enable each Party (and their respective Affiliates) to comply with its legal and regulatory obligations. Unless otherwise agreed by the Parties, the Pharmacovigilance Agreement will assign responsibility for maintaining the global safety database for the Product [***], and will provide that all costs associated with maintaining the global safety database shall be

considered Other Shared Expenses. The Pharmacovigilance Agreement will contain terms no less stringent than those required by ICH or other applicable guidelines in order to allow the Parties to meet the applicable regulatory and legal requirements regarding the management of safety data in their respective territories. Each Party shall perform its pharmacovigilance obligations in connection with the Product in compliance with the Pharmacovigilance Agreement and all Applicable Laws.

7.6.2 **Pharmacovigilance Audit**. In accordance with timing mutually agreed to by the Parties, but in any event prior to the expected initiation of the first clinical trial for the Product in the Kiniksa Field, Regeneron may conduct a one-time pharmacovigilance qualification and capability audit of Kiniksa or its designee to ensure that Kiniksa or such designee can perform safety and pharmacovigilance reporting activities in accordance with Applicable Laws. Kiniksa shall not initiate any clinical trials for the Product unless such audit is reasonably satisfactory to Regeneron. In accordance with timing mutually agreed to by the Parties, but in any event prior to the expected transfer of the global safety database in accordance with Section 3.3.2 (Regulatory and Safety Information), Regeneron shall conduct a one-time pharmacovigilance qualification and capability audit of Kiniksa to ensure that Kiniksa can perform safety and pharmacovigilance reporting activities as the holder of the global safety database for the Product in accordance with Applicable Law. On a periodic basis, but no more than once per Contract Year, on [***] days' advance written notice, Regeneron may conduct ongoing pharmacovigilance capability audits to ensure Kiniksa is meeting its obligations under the Pharmacovigilance Agreement. Kiniksa shall consider in good faith any comments made by Regeneron in connection with any audit conducted pursuant to this Section 7.6.2 (Pharmacovigilance Audit). Each Party's reasonable Fully-Burdened Costs incurred in connection with any audit conducted pursuant to this Section 7.6.2 (Pharmacovigilance Audit) shall be considered Other Shared Expenses, except that Regeneron shall bear its own costs for any ongoing periodic audits, unless Regeneron conducts such audits for good cause (*e.g.*, Kiniksa receives a warning letter from a Regulatory Authority) or such audit determines that Kiniksa failed to meet its obligations under the Pharmacovigilance Agreement.

7.7 **Regulatory Inspection or Audit**. If a Regulatory Authority desires to conduct an inspection or audit of a Party with regard to any Product intended for use in the Kiniksa Field in the Territory (excluding a general facilities inspection or audit), then each Party agrees to reasonably cooperate with the other Party and such Regulatory Authority during such inspection or audit, including by allowing, to the extent practicable, a representative of the other Party to be present during the applicable portions of such inspection or audit to the extent it relates to the Development, Commercialization, or Manufacturing of any Product in the Territory. Following receipt of the inspection or audit observations of the Regulatory Authority (a copy of which the receiving Party will promptly provide to the other Party), the Party in receipt of the observations will prepare any appropriate responses; provided that the other Party, to the extent practicable, will have the right to review and comment on such responses to the extent they relate to, or may be reasonably expected to adversely impact, any Product in the Kiniksa Field in the Territory, and the Party in receipt of the observations will consider in good faith the comments made by the other Party within [***] days of receipt by such other Party, or such shorter period as may be required to satisfy requirements under Applicable Law. In the event the Parties disagree concerning the form or content of a response, then the Party that received the observations will decide the appropriate form and content of the response. Without limiting the foregoing, each Party (and its Third Party subcontractors) must, to the extent reasonably possible, notify the other Party no later than [***] after receipt of a notification from a Regulatory Authority of the intention of such Regulatory Authority to audit or inspect facilities used or proposed to be used for the Manufacture of any Product under this Agreement; provided that, to the extent reasonably possible, such notification must be given to the other Party no later than [***] prior to any such Regulatory Authority audit or inspection. In the event that the Party that is subject to such audit or inspection by a Regulatory Authority is not provided with advance notice thereof, then such Party will inform the other Party of the occurrence thereof within [***]. The Fully-Burdened Cost incurred by or on behalf of Regeneron in good faith in the course of any audit or inspection by a Regulatory Authority shall be considered Other Shared Expenses.

7.8 **Recalls and Other Corrective Actions**. If a Party believes that a recall or market withdrawal of a Product in the Territory may be required, then such Party will so notify the other Party within [***], and, without limitation of and subject to this Section 7.8 (Recalls and Other Corrective Actions), the Parties will decide whether or not such a recall or market withdrawal is required. Decisions with respect to any recall, market withdrawal, or other corrective action related to any Product in the Kiniksa Field in the Territory will be made [***]; provided, however, that if the [***] will have such final decision authority on and after the U.S. Marketing Approval (the Party responsible for recalls or market withdrawals pursuant to this Section 7.8 (Recalls and Other Corrective Actions), the "Recall Responsible Party"). Notwithstanding the foregoing (including a Party's final decision making authority), nothing herein will prohibit either Party from initiating or conducting any recall or other corrective action mandated by any Governmental Authority or Applicable Law. If the non-Recall Responsible Party wishes to initiate a recall or market withdrawal of a Product in the Territory in good faith, but the Parties are unable to agree on whether or not to initiate any such recall or other action,

and thereafter the Recall Responsible Party does not initiate a recall or market withdrawal of a Product in the Territory, then [***]. The Parties will cooperate with respect to any actions taken or public statements made in connection with any such recall or market withdrawal. Any recall of any Product in the Territory caused by [***]. Any recall caused by [***]. Any recall of the Product in the Kiniksa Field in the Territory not [***].

ARTICLE 8
MANUFACTURING AND SUPPLY

8.1 **Regeneron Supply of Product.**

8.1.1 **Exclusive Right for Regeneron to Supply Product**. Until the occurrence of a Manufacturing Technology Transfer Event, Regeneron shall have the exclusive right to Manufacture or have Manufactured and supply or have supplied the Product for all Development and Commercialization purposes in the Kiniksa Field in the Territory. As of the Effective Date, Regeneron or one of its Affiliates (a) Manufactures the Formulated Bulk Product in its own facilities, and (b) uses a Third Party Fill/Finish Provider to perform the finishing, filling, assembly, packaging, and testing of all Filled Product.

8.1.2 **Reestablishing Protocols**. The Parties acknowledge and agree that Regeneron will need to reestablish protocols for the Manufacture of the 80mg dosage form for the Existing Product Formulation if such dosage form is requested by Kiniksa under the Supply Agreements. Regeneron's Fully-Burdened Cost for the performance of activities related to re-establishing the protocols for the Manufacture of the 80mg dosage form of the Existing Product Formulation shall be considered Other Shared Expenses.

8.1.3 **Notification of Transfer of Manufacturing Activities**. If at any time during the Term, Regeneron desires to transfer a portion or all of the Manufacturing activities for the Product to any of its Affiliates or one or more Third Parties, then Regeneron will first provide written notice thereof to Kiniksa in accordance with the Quality Agreement; *provided that*, consistent with Section 8.14.1 (Manufacturing Technology Transfer Event), Regeneron may not provide notice that it (or its Affiliates) wishes to discontinue the Manufacture of any Formulated Bulk Product under the Supply Agreement prior to the earlier of (a) [***] years after the date of receipt of U.S. Marketing Approval, or (b) [***] after the Effective Date, if U.S. Marketing Approval has not been obtained. If at any time during the Term, Regeneron desires to transfer a portion or all of such Manufacturing activities to one or more Third Parties, then Regeneron shall consider in good faith any Third Party contract manufacturers proposed by Kiniksa. After U.S. Marketing Approval, at Kiniksa's request, the Parties shall discuss whether Kiniksa (instead of Regeneron) should directly enter into a supply agreement with the agreed Third Party contract manufacturer if Regeneron delivers notice that Regeneron desires to transfer a portion or all of such Manufacturing activities to one or more Third Parties. Regeneron shall

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remain responsible under this Agreement for the performance of any such Affiliates or Third Parties and Regeneron will be responsible for any technology transfer activities that may be required to enable such Affiliates or Third Party to perform such Manufacturing activities.

8.1.4 **Decisions Regarding Manufacturing Transfer**. The Parties will consult and cooperate with each other with respect to any such proposed transfer of all or a portion of such Manufacturing activities and attempt to reach all such decisions by consensus, but, notwithstanding anything to the contrary in Section 8.1.3 (Notification of Transfer of Manufacturing Activities), (a) [***], (b) [***], and (c) [***]. The Party that is the holder of the U.S. BLA will have responsibility for, and final decision making with respect to, filing such any and all Registration Filings required by the FDA or under Applicable Law in connection with any such transfer of Manufacturing activities; *provided that* the Parties will consult and cooperate with each other regarding such Registration Filings.

8.1.5 **Continuation of Supply**. In the case of any transfer by Regeneron of Manufacturing pursuant to Section 8.1.3 (Notification of Transfer of Manufacturing Activities) or in connection with any Manufacturing Technology Transfer Event, Regeneron will use Commercially Reasonable Efforts to continue to Manufacture or have Manufactured and supply or have supplied the Product for all Development and Commercialization purposes in the Kiniksa Field in the Territory in accordance with the applicable Supply Agreement for a period of [***] (unless a shorter period of time is requested by Kiniksa) after (a) Regeneron's decision to transfer Manufacturing pursuant to Section 8.1.3 (Notification of Transfer of Manufacturing Activities), or (b) the occurrence of a Manufacturing Technology Transfer Event and Regeneron's consent to the Third Party contract Manufacturer proposed by Kiniksa, as set forth in Section 8.15.3 (Consent). Notwithstanding the foregoing, Regeneron shall not unreasonably withhold, condition, or delay its consent to extend such time period, if a reasonable extension is requested in good faith by Kiniksa if such Manufacturing Technology Transfer is not complete [***].

8.2 **Development Supply**. Regeneron shall use Commercially Reasonable Efforts to Manufacture (or have Manufactured) and supply to Kiniksa (or have supplied) all Product and placebo necessary to conduct Development in accordance with the terms and conditions of the Development Supply Agreement.

8.3 **Development Supply Agreement**. Within [***] following the Effective Date (or such other timeframe as may be agreed by the Parties), the Parties shall negotiate and enter into a definitive supply agreement for Regeneron's supply to Kiniksa of its Clinical Supply Requirements solely for use in conducting Development activities ("Development Supply Agreement"). The Development Supply Agreement shall provide for customary terms and conditions, including forecasting, ordering, delivery, payment, and supply, and

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shall be consistent with the terms set forth in Schedule 8.3 and the terms of this Agreement. Regeneron may designate an Affiliate to enter into the Development Supply Agreement.

8.4 **Commercial Supply**. Regeneron shall use Commercially Reasonable Efforts to Manufacture (or have Manufactured) and supply to Kiniksa (or have supplied) all Product necessary to conduct Commercialization in the Kiniksa Field in accordance with the Commercial Supply Agreement.

8.5 **Commercial Supply Agreement**. At least [***] prior to the anticipated date of receipt of U.S. Marketing Approval, the Parties shall negotiate a definitive commercial supply agreement (the "Commercial Supply Agreement", and together with the Development Supply Agreement, the "Supply Agreements") for the supply of the Commercial Supply Requirements to Kiniksa in accordance with the Commercial Plan. The Commercial Supply Agreement shall provide for customary terms, including with respect to forecasting, ordering, delivery, payment, and supply, and the terms of this Agreement. Regeneron may designate an Affiliate to enter into the Commercial Supply Agreement. Except as otherwise agreed by the Parties and set forth in the Commercial Supply Agreement, all Product supplied under the Commercial Supply Agreement shall be in the form of Formulated Bulk Product or Filled Product, as such form is determined by Kiniksa, except that after a Fill/Finish Technology Transfer requested by Kiniksa in accordance with Section 8.14.2 (Fill/Finish Transfer), the Product shall be supplied in the form of Formulated Bulk Product. At Kiniksa's request, to the extent Product is supplied in Filled Product form, Regeneron shall request that its Third Party Fill/Finish Provider package and label such Filled Product in accordance with Kiniksa's instructions and shall use reasonable efforts to facilitate the transfer of Kiniksa's packaging and labeling instructions to such Third Party Fill/Finish Provider. The Parties acknowledge and agree that as of the Effective Date, the Third Party Fill/Finish Provider's order requirements are for a minimum of [***] vials of the Product per purchase order.

8.6 **Price for Product Supplied by Regeneron**. The price for Product supplied by Regeneron to Kiniksa under the Supply Agreements shall be the [***] during a period, as set forth in [***] for such period, [***]. Regeneron shall invoice Kiniksa for the [***] concurrently with the delivery of the Product to Kiniksa, unless otherwise agreed by the Parties.

8.7 **Quality Agreements**. No later than [***] following the Effective Date (or such other timeframe as may be agreed by the Parties), but in any event prior to the commencement of the first human clinical trial for any Product to be conducted by Kiniksa in the Kiniksa Field in the Territory, the Parties shall negotiate and enter into a reasonable and customary quality agreement with respect to the Product to be Manufactured by or on behalf of Regeneron and supplied to Kiniksa under the Development Supply Agreement (the "Development Quality Agreement"). At least [***] prior to the anticipated date of receipt of U.S. Marketing Approval, the Parties shall negotiate and enter into a reasonable and customary quality agreement with respect to the Product to be Manufactured by or on

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behalf of Regeneron and supplied to Kiniksa under the Commercial Supply Agreement (the "Commercial Quality Agreement" and together with the Development Quality Agreement, the "Quality Agreements").

8.8 **Product Changes**.

8.8.1 **Product Change Required under Applicable Law in the Territory**. All Product Changes shall be made in accordance with the Quality Agreements. So long as Regeneron is Manufacturing the Product under a Supply Agreement, Regeneron shall use Commercially Reasonable Efforts to implement (or have implemented) any Product Changes required under Applicable Law in the Territory.

8.8.2 **Other Product Changes Requested by Kiniksa**. Regeneron shall use Commercially Reasonable Efforts to implement (or have implemented) all Product Changes requested by Kiniksa to the extent approved by Regeneron, such approval not to be unreasonably withheld, conditioned, or delayed.

8.8.3 **Registration Filings**. Prior the U.S. BLA Transfer Date, Regeneron shall prepare any necessary supplementary Registration Filings for the Product in the Kiniksa Field in the Territory with respect to any Product Changes and provide such Registration Filings to Kiniksa for its review and comment reasonably in advance of the filing thereof (and use good faith efforts to incorporate any such comments), and Regeneron shall be responsible for filing such Registration Filings with respect to such Product Changes. After the U.S. BLA Transfer Date, Kiniksa, in consultation with Regeneron, shall prepare any necessary supplementary Registration Filings for the Product in the Kiniksa Field in the Territory with respect to any Product Changes, and Kiniksa shall be responsible for filing such Registration Filings with respect to any such Product Changes and shall file any updates to the U.S. BLA that may be associated with any such Product Changes. After the U.S. BLA Transfer Date, Kiniksa shall promptly notify and provide Regeneron with copies of any updates to the U.S. BLA.

8.8.4 **Costs of Product Changes**. The Fully-Burdened Cost incurred by or on behalf of Regeneron in connection with any activities directly related to any (a) Product Changes required by under Applicable Law in the Territory (except for Product Changes required under Applicable Law that exclusively relate to the Regeneron Field), (b) any discretionary Product Changes requested by Kiniksa and approved in advance in writing by Regeneron, such consent not to be unreasonably, withheld, conditioned, or delayed, or (c) discretionary Product Changes made by Regeneron but only to the extent approved in writing in advance by Kiniksa, such consent not to be unreasonably, withheld, conditioned, or delayed, and that such Product Changes benefit a Product (*e.g.*, changes that reduce Manufacturing Cost), in each case ((a) through (c)), shall be considered Other Shared Expenses (and, with respect to Product Changes set forth in clause (c), if consent is not obtained from Kiniksa, then the cost of implementing such

Product Changes shall not be considered Other Shared Expenses). Notwithstanding the foregoing, if any such Product Changes in clause (c) are applicable to Products in both the Kiniksa Field and in the Regeneron Field, then, prior to the receipt of U.S. Marketing Approval, [***], unless otherwise agreed to by the Parties. If Kiniksa incurs any Out-of-Pocket Costs with respect to filing any updates to the U.S. BLA associated with any Product Changes after the U.S. BLA Transfer Date, then Kiniksa's Fully-Burdened Cost for the performance of activities under this Section 8.8 (Product Changes) shall be considered Other Shared Expenses, other than with respect to Product Changes that are exclusively related to the Retained Field or that are discretionary Product Changes made by Regeneron that do not benefit a Product (*e.g.*, changes made by Regeneron for internal space planning purposes), in which case, [***] of Kiniksa in connection with the performance of activities related to such Product Changes.

8.9 **Manufacturing Compliance**. Regeneron shall use Commercially Reasonable Efforts to Manufacture the Product under each Supply Agreement, and as applicable ensure that all Products are Manufactured by Third Parties, in each case, in conformity with Good Practices, and the Quality Agreements, including the Product Specifications and Applicable Law.

8.10 [***]. Except to the extent specifically required by Kiniksa or required by a Regulatory Authority in the Kiniksa Field in the Territory with respect to any Product, any [***] the Manufacture of Products shall be the [***].

8.11 **Defective Product.**

8.11.1 Limitation on Damages. Without limiting Section 14.2.6 (Indemnity by Regeneron), except for Regeneron's liability for recalls and withdrawals as set forth in Section 7.8 (Recalls or Other Corrective Actions), Kiniksa's sole and exclusive remedy, and Regeneron's (or its Affiliate's) sole obligation and liability, with respect to defective or Non-Conforming Products or failure to supply in accordance with the terms of the applicable Supply Agreement shall be (i) [***], (ii) [***]. For clarity, nothing in this Section 8.11.1 (Limitation of Damages) shall limit Section 14.2.6 (Indemnity by Regeneron).

8.11.2 **Non-Conforming Product**. If the Parties are unable to agree as to whether any Product is Non-Conforming Product, or to determine the cause of any Non-Conforming Product, then, in each case, an outside laboratory or consultant reasonably agreeable to each Party will conduct an investigation to determine the cause of any alleged Non-Conforming Product. Such outside testing laboratory or consultant will determine, using samples of the applicable lot of Product, whether the lot of Product is Non-Conforming Product and the cause of any non-conformity, if able to do so. The test results obtained from such laboratory and the determinations of such laboratory shall be final and binding upon each Party with respect to conformity or Non-Conforming Product and the

cause of any such non-conformity. The fees and expenses of such testing shall be considered Other Shared Expenses.

8.11.3 **Latent Defects, Adulteration or Misbranding**. If, after delivery in any Product supplied by Regeneron, either Party finds a previously undetected non-conformity to the Product or that such Product was adulterated or misbranded, the discovering Party shall promptly notify the other Party.

8.12 **Risk Mitigation**. Upon execution of the Development Supply Agreement, with input and advice from Regeneron, Kiniksa shall develop a risk mitigation plan and supply chain strategy intended to ensure continuous uninterrupted supply of Product. With input and advice from Regeneron, Kiniksa shall update such plan upon execution of the Commercial Supply Agreement, and from time-to-time as may be necessary during the Term. In addition, at all times during the Term, Regeneron will ensure that it maintains supplies of raw materials sufficient to Manufacture quantities of the Product in accordance with any firm order requirements of Kiniksa in the Supply Agreements.

8.13 **Notification and Discussion of Supply Issues**. If Regeneron (a) reasonably determines that it will not be able to supply any Product in accordance the Development Supply Agreement or Commercial Supply Agreement, or (b) becomes aware of any problems or delays of any material nature in performing its contractual obligations that have the potential to adversely affect the Development or Commercialization of any Product, as the case may be (each, an "Anticipated Supply Shortage"), then Regeneron shall provide written notice to Kiniksa of such Anticipated Supply Shortage within [***] of the date Regeneron becomes aware of the same, together with a detailed explanation of the reasons related to such Anticipated Supply Shortage and of the expected duration thereof. Regeneron will keep Kiniksa informed on a timely basis of any developments during any such Anticipated Supply Shortage. Upon Regeneron's notification to Kiniksa regarding any Anticipated Supply Shortage, subject to and without limiting any applicable terms and conditions in any Supply Agreement or this Agreement, the JSC will discuss such shortage pursuant to Section 4.1.3(m) (Duties of the JSC), and determine if an alternate Third Party supply source should be utilized, and the identity of any such Third Party supply source, if so determined by the JSC. [***].

8.14 **Manufacturing Technology Transfer Event**. The occurrence of any of the following events shall be a "Manufacturing Technology Transfer Event" for purposes of this Agreement.

8.14.1 **Discontinuation of Formulated Product Manufacturing**. In the event that Regeneron determines that it (or its Affiliates) desires to discontinue the Manufacture of any Formulated Bulk Product under a Supply Agreement; *provided that* Regeneron shall provide Kiniksa

with prior written notice of such discontinuance to enable the Parties to effectuate a complete and successful Manufacturing Technology Transfer, and with respect to Formulated Bulk Product, such notice may not be provided prior to the earlier of (i) [***], or (ii) [***]. If Regeneron provides notice pursuant to this Section 8.14.1

(Discontinuation of Manufacturing), then the terms of Section 8.1.5 (Continuation of Supply) shall apply during the period after Regeneron's delivery of such notice;

8.14.2 **Fill/Finish Transfer**.

(a) Prior to U.S. Marketing Approval. Prior to the date of U.S. Marketing Approval, if Kiniksa notifies Regeneron in writing that it wishes to receive a Manufacturing Technology Transfer for the fill/finish process to convert the Formulated Bulk Product into Filled Product (a "Fill/Finish Technology Transfer"), then Regeneron shall consider such request in good faith; and

(b) After U.S. Marketing Approval. At any time after the date of U.S. Marketing Approval, if Kiniksa notifies Regeneron in writing that it wishes to receive a Fill/Finish Transfer, then Regeneron will perform such Fill/Finish Technology Transfer in accordance with timelines agreed by the Parties;

8.14.3 **Anticipated Supply Shortage**. If the JSC is unable to resolve any Anticipated Supply Shortage, and based on forecasts provided by Kiniksa under the Supply Agreements and Regeneron's production schedule for such Product, Regeneron will not be able to meet at least [***] of Kiniksa's Product forecast over a period of [***].

8.14.4 **Failure to Supply**. During any [***] period, Regeneron's failure to supply in accordance with Section 8.9 (Manufacturing Compliance) conforming Product with respect to [***] of the aggregate quantity of all Products ordered pursuant to binding purchase orders on or before the firm delivery dates in the applicable binding purchase orders delivered in accordance with the Supply Agreements.

8.15 **Manufacturing Technology Transfer**. Upon the occurrence of a Manufacturing Technology Transfer Event, the following shall occur:

8.15.1 **Safety Stock**. From and after the date of notice by Regeneron to Kiniksa of a Manufacturing Technology Transfer Event pursuant to Section 8.14.1 (Discontinuation of Formulated Product Manufacturing), in accordance with the terms of the applicable Supply Agreement, Kiniksa may order additional quantities of Product under the applicable Supply Agreement to hold as safety stock.

8.15.2 **Transfers**. Regeneron shall use reasonable efforts to transfer (or have transferred) the Manufacture of the Formulated Bulk Product or Filled Product to the facility designated for the Manufacturing Technology Transfer in accordance with Section 8.1.3 (Notification of Transfer of Manufacturing Activities), including making available to Kiniksa or its designee all Regeneron

Manufacturing Know-How by providing to Kiniksa (or its designee) all Product Manufacturing Records, in each case, related to Products in the Kiniksa Field (including for clarity, those Product Manufacturing Records related to the Regeneron Field if such transfer occurs after the date of receipt of U.S. Marketing Approval) and copies or samples of relevant documentation, materials, and other embodiments of such Know-How (with respect to Formulated Bulk Product, a "Formulated Bulk Technology Transfer" and together with a Fill/Finish Technology Transfer, each a "Manufacturing Technology Transfer"). Subject to Section 8.1.5 (Continuation of Supply), each Manufacturing Technology Transfer shall be commenced within a commercially reasonable timeframe and conducted pursuant to a technology transfer plan to be promptly developed and agreed by the Parties (each, a "Technology Transfer Plan") for the purpose of ensuring the complete and timely transfer of such Regeneron Manufacturing Know-How in a manner that is consistent with Regeneron's then-current internal technology transfer corporate standards. In connection with such Manufacturing Technology Transfer, Regeneron shall transfer working cell banks to Kiniksa or its designee. Regeneron shall transfer additional quantities of working cell banks to Kiniksa to the extent reasonably required by Kiniksa from time to time; *provided that* nothing in this Section 8.15 (Manufacturing Technology Transfer) shall require Regeneron to transfer any portion of the master cell bank to Kiniksa. In addition, at Kiniksa's request, Regeneron will use reasonable efforts (which efforts will not, unless otherwise agreed by the Parties, include any obligation to make any payment or incur economic burden) to facilitate a direct relationship between Kiniksa and Regeneron's Third Party Fill/Finish Provider. Regeneron shall [***]. In addition, at Regeneron's request, Kiniksa will use reasonable efforts (which efforts will not, unless otherwise agreed by the Parties, include any obligation to make any payment or incur economic burden) to discuss entering into a direct relationship with Neovii under which Kiniksa would Manufacture (or have Manufactured) and supply (or have supplied) the Product for Neovii (to the extent such Manufacturing activities are subject to the Manufacturing Technology Transfer).

8.15.3 **Consent**. Kiniksa's request to designate a Third Party contract Manufacturer shall be subject to Regeneron's prior written consent as set forth in Section 2.7.2 (Sublicensing by Kiniksa); *provided that* such consent shall not be unreasonably withheld, conditioned, or delayed,

and the [***] period set forth in Section 8.1.5 (Continuation of Supply) during which Regeneron will continue to Manufacture or have Manufactured and supply or have supplied the Product for all Development and Commercialization purposes in the Kiniksa Field in the Territory will not commence until the time at which Regeneron gives its consent to such a Third Party contract Manufacturer proposed by Kiniksa.

8.15.4 **Lonza License**. [***]. Upon the occurrence of a Manufacturing Technology Transfer Event, Regeneron shall use reasonable efforts (which efforts will not,

unless otherwise agreed by the Parties, include any obligation to make any payment or incur economic burden) [***] for Kiniksa or its Third Party designee to Manufacture the applicable Products, and the [***] period set forth in Section 8.1.5 (Continuation of Supply) during which Regeneron will continue to Manufacture or have Manufactured and supply or have supplied the Product for all Development and Commercialization purposes in the Kiniksa Field in the Territory will be tolled until the time [***].

8.15.5 **Assistance and Observation**. In connection with any Manufacturing Technology Transfer Regeneron shall provide Kiniksa with reasonable technical consultation and assistance (including answering reasonable questions and making appropriate personnel reasonably available) regarding the transferred Manufacturing processes to facilitate the success transition of the Manufacture of the Product to Kiniksa or its designee. To the extent such Manufacturing is performed by Regeneron, Regeneron shall allow an agreed number of representatives of Kiniksa to observe the Manufacturing processes in otherwise scheduled manufacturing runs (but without any obligation to conduct a manufacturing run for purposes of such observation, and subject to customary restrictions and obligations applicable to visitors), all in accordance with the Technology Transfer Plan.

8.15.6 **Manufacturing Technology Transfer Costs**. Kiniksa shall [***]. For clarity, Kiniksa's Fully-Burdened Costs incurred connection with a Fill/Finish Technology Transfer undertaken in accordance with Section 8.14.2 (Fill/Finish Transfer) (and not in connection with Formulated Bulk Technology Transfer), [***].

ARTICLE 9
PAYMENTS

9.1 **Upfront Payment**. Within five (5) days after the Effective Date, Kiniksa shall pay to Regeneron a non-refundable, non-creditable amount of Five Million Dollars (US$5,000,000).

9.2 **Development Milestones**.

9.2.1 **Milestone Payments**. Kiniksa shall inform Regeneron in writing within [***] after the occurrence of a Development Milestone Event. Kiniksa shall make a one-time, non-refundable, non-creditable milestone payment to Regeneron within [***] days from receipt of an invoice from Regeneron, such invoice to be provided to Kiniksa after Kiniksa's notice to Regeneron of the first achievement of each of the milestone events set forth in Table 9.2 below for the first Product (each, a "Development Milestone Event" and the corresponding payment set forth in Table 9.2, a "Development Milestone Payment").

TABLE 9.2 — Development Milestones

	Development Milestone Event	Development Milestone Payment ($)
(A)	[***]	[***]
(B)	[***]	[***]

9.2.2 **Achievement of Development Milestones**. Each of the Development Milestone Payments set forth in Table 9.2 (Development Milestones) above is payable only once. No Development Milestone Payments shall be payable for any subsequent Product to achieve such Development Milestone Event. The maximum amount payable by Kiniksa in respect of Development Milestone Payments if all Development Milestones Events occur will be $27,500,000.

9.3 **Sharing of Third Party Proceeds**. The Parties shall share Third Party Proceeds in accordance with their Regeneron Third Party Proceed Percentages. Third Party Proceeds shall be shared when received by Kiniksa or its Affiliates on a cash basis and shall be calculated independently and shall not be offset by any calculation of Profits or the Regeneron Profit Percentage, as further illustrated in the example set forth on Schedule 9.4.

9.4 **Sharing of Profits, Periodic Reports, and Fund Flow Mechanics**. The Parties shall share Profits in accordance with their Regeneron Profit Percentages as described in this Section 9.4 (Sharing of Profits) and as further illustrated in the example set forth on Schedule 9.4.

9.4.1 **Periodic Reports**. Kiniksa and Regeneron shall each prepare and deliver to the other Party the periodic reports specified in this Section 9.4.1 (Periodic Reports). All reports referred to in this Section 9.4.1 (Periodic Reports) shall be in such form, format, and level of detail approved by the JSC pursuant to Section 4.1.3(n) (Duties of the JSC). Unless otherwise agreed by the JSC, the financial data in the reports to be provided hereunder will include calculations in United States Dollars. As disagreement between the Parties regarding any amount included in the Profit Payment Report, Third Party Proceeds Payment Report, or in any Quarterly Expense Report, will, in each case, be a Financial Dispute that is resolved in accordance with Section 17.1.2 (Financial Disputes).

(a) Quarterly Net Sales. Within [***] following the end of each Quarter (and within [***] following the end of the fourth (4th) Quarter of each Contract Year), commencing with the Quarter in which Kiniksa or its Affiliates receives Net Sales in any country in the Territory, Kiniksa shall deliver electronically to Regeneron a written report setting forth, on

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a country-by-country basis for such Quarter, for each country in the Territory, (i) the Net Sales invoiced by Kiniksa or its Affiliates during such Quarter in such country, (ii) quantities of the Product sold in such country in such Quarter, and (iii) gross Product sales in such country in such Quarter, along with an accounting of the deductions from gross sales permitted by the definition of Net Sales. In addition, within [***] following the end of each Quarter, commencing with the Quarter in which Kiniksa or its Affiliates receives Net Sales in any country in the Territory, Kiniksa shall deliver electronically to Regeneron a written "flash" report setting forth, on a country-by-country basis for such Quarter, for each country in the Territory, Kiniksa's good faith estimate of the amounts set forth in the foregoing clauses (i) through (iii). To the extent any sales were attributable to erroneous or reversed invoices by Kiniksa or its Affiliates during a Quarter and included in the report for a Quarter pursuant to this Section 9.4.1(a) (Quarterly Net Sales) (other than bad debt pursuant to Section 1.189(h) (Shared Commercial Expenses)) and only recognized after Kiniksa delivered the report for such Quarter pursuant to this Section 9.4.1(a) (Quarterly Net Sales), Kiniksa shall be entitled to deduct such amounts in the Quarter following the Quarter in which such sales were mistakenly booked.

(b) Quarterly Incurred Expenses. Within [***] following the end of each Quarter (and, with respect to Quarterly Expense Reports to be provided by Kiniksa, within [***] following the end of the fourth (4th) Quarter in each year), each Party that has incurred any Other Shared Expenses, Shared Commercial Expenses, Cost of Goods Sold, or Manufacturing Technology Transfer Costs in that Quarter shall deliver electronically to the other Party a written report setting forth in reasonable detail such Other Shared Expenses, Shared Commercial Expenses, or Cost of Goods Sold, in each case, incurred by such Party on a country-by-country basis in such Quarter in the Territory (the "Quarterly Expense Report"). In addition, within [***] following the end of each Quarter, each Party that has incurred any Other Shared Expenses, Shared Commercial Expenses, Cost of Goods Sold, or Manufacturing Technology Transfer Costs in that Quarter shall deliver electronically to the other Party a written "flash" report setting forth such Party's good faith estimate of the amounts to be included in each Quarterly Expense Report.

(c) Quarterly Profit Payment Report. Within [***] following the end of each Quarter (and within [***] following the end of the fourth (4th) Quarter in each year), Kiniksa shall deliver electronically to Regeneron a Profit Payment Report in respect of such Quarter, combining the information reported by each Party as Net Sales pursuant to Section 9.4.1(a) (Quarterly Net Sales) and in the Quarterly Expense Report pursuant to Section 9.4.1(b) (Quarterly Incurred Expenses) and showing its

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calculations in accordance with Schedule 9.4 of the amount of any Regeneron Profit Split for such Quarter as contemplated by this Section 9.4.1(c) (Quarterly Profit Payment Report) (including showing the calculation of such Regeneron Profit Split). In addition, within [***] following the end of each Quarter, Kiniksa shall deliver electronically to Regeneron a written "flash" report setting forth Kiniksa's good faith estimate of the amount of any Regeneron Profit Split for such Quarter. For clarity, if an item is included in a Profit Payment Report for a Quarter, in no event shall the same item be included in a subsequent Profit Payment Report.

(d) Quarterly Report of Third Party Proceeds. Within [***] following the end of each Quarter in which Kiniksa receives Third Party Proceeds, Kiniksa shall deliver electronically to Regeneron a report in respect of such Quarter, providing information regarding the amount of Third Party Proceeds, the identity of the Third Party, and the characterization and calculation of the Regeneron Third Party Proceeds Split ("Third Party Proceeds Payment Report"). For clarity, the Third Party Proceeds Payment Report shall be calculated independently and not reduced by any expenses in a Profit Payment Report. In addition, within [***] following the end of each Quarter in which Kiniksa receives any Third Party Proceeds, Kiniksa shall deliver electronically to Regeneron a

written "flash" report setting forth Kiniksa's good faith estimate of the amounts to be included in each Third Party Proceeds Payment Report.

(e) Quarterly Inventory Report. Within [***] days after the end of each Quarter (for the first three (3) Quarters in a Contract Year), commencing with the Quarter in which the First Commercial Sale of the Product in the Kiniksa Field in the Territory occurs, Kiniksa shall deliver electronically to Regeneron a written inventory report (the "Quarterly Inventory Report") setting forth the ending inventory of the Product balance for that Quarter. To the extent reasonably practicable, each Quarterly Inventory Report shall provide such information broken out by lot numbers, dosage form, and unit size, for units of Product contained in such report.

(f) Annual Inventory Report. Within [***] after the end of each Contract Year, commencing with the Quarter in which the First Commercial Sale of the Product in the Kiniksa Field in the Territory occurs, Kiniksa shall deliver electronically to Regeneron a written inventory report (the "Annual Inventory Report") reconciling beginning and ending inventory and including (i) the number of units of Product distributed, but not sold (such as samples, donations, and write-offs) in the Territory during such Contract Year, (ii) the number of units of Product for the Territory that are lost, destroyed, expired, or become obsolete or spoiled during such

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Contract Year, and (iii) the number of units of Product sold in the Territory during such Contract Year. To the extent reasonably practicable, each Annual Inventory Report shall provide such information broken out by lot numbers, dosage form and unit size, for units of Product contained in such report.

(g) Flash Reports. Each Party acknowledges and agrees that the "flash reports" to be provided under this Section 9.4.1 (Periodic Reports) represent only the applicable Party's good faith estimate of the costs, expenses, or profits (as applicable) to be provided in the applicable report with the use of Commercially Reasonable Efforts. Neither Party will be responsible in connection with any inaccuracies included in any flash report provided pursuant to this Section 9.4 (Sharing of Profits) to the extent a Party complied with the previous sentence. Each Party acknowledges and agrees that the "flash reports" to be provided under this Section 9.4.1 (Periodic Reports) (rather than having the definitive reports due at such time period) is an accommodation to Kiniksa while it is a young company. As Kiniksa develops its accounting and financial reporting capabilities, the Parties shall discuss in good faith, the shortening of the time periods in Section 9.4.1 (Periodic Reports) and any related changes in Section 9.4.5 (Funds Flow).

9.4.2 **Reimbursement of Manufacturing Costs**.

(a) Kiniksa Payment to Regeneron for Product Supplied under the Supply Agreement. Within [***] following the end of each Quarter in which Regeneron supplies Kiniksa with Product under the Supply Agreements, or is otherwise entitled to invoice Kiniksa for Product to be supplied under the Supply Agreements (if earlier), Regeneron shall deliver electronically to Kiniksa a written report setting forth the [***] (as set forth in Section 8.6 (Price for Product Supplied by Regeneron)), [***].

(b) [***]. Within [***] following the end of each Contract Year in which Regeneron supplies Kiniksa with Product under the Supply Agreements, Regeneron shall deliver electronically to Kiniksa a written report setting forth [***] and [***] and the [***] (the "[***] Amount").

(c) [***] Amount. [***] invoiced by Regeneron pursuant to Section 9.4.2(a) (Kiniksa Payment to Regeneron for Product Supplied under the Supply Agreement) for Product supplied by Regeneron for a Contract Year [***] for Product supplied by Regeneron for a Contract Year, [***] (Payments of Certain Invoiced Amounts). [***] (Kiniksa Payment to Regeneron for Product Supplied under the Supply Agreement) for Product supplied by Regeneron for a Contract Year [***].

9.4.3 **Kiniksa Incurred Expenses under Section 8.8.4 (Cost of Product Changes)**. Within [***] following the end of each Quarter in which Kiniksa incurs any Out-of-Pocket Expenses with respect to filing any updates to the U.S. BLA associated with any Product Changes after the U.S. BLA Transfer Date for which Kiniksa is entitled to reimbursement from Regeneron as set forth Section 8.8.3 (Cost of Product Changes), Kiniksa shall deliver electronically to Regeneron a written report setting forth in reasonable detail such actual Out-of-Pocket Expenses in such Quarter. In addition, within [***] days following the end of each Quarter in which Kiniksa incurs any Out-of-Pocket Expenses with respect to filing any updates to the U.S. BLA associated with any Product Changes after the U.S. BLA Transfer Date, Kiniksa shall deliver electronically to Regeneron a written "flash" report setting forth Kiniksa's good faith estimate of such amounts.

9.4.4 **Other Shared Expenses**. Notwithstanding anything to the contrary set forth herein, in no event shall Regeneron request reimbursement for any expenses that would be classified as Other Shared Expenses under this Agreement or that are otherwise due to Regeneron hereunder, in each case, to the extent Neovii has reimbursed or is reimbursing Regeneron for such expenses under the Neovii License Agreement.

9.4.5 **Funds Flow**. For each Quarter:

(a) Regeneron Expenses. Kiniksa [***] for such Quarter in its Quarterly Expense Report provided pursuant to Section 9.4.1(b) (Quarterly Incurred Expenses) in accordance with Section 9.4.5(c) (Payment to Regeneron).

(b) Kiniksa Expenses. Kiniksa will [***] for such Quarter reflected in the Profit Payment Report for such Quarter.

(c) Payment to Regeneron of Profits. Prior to the first receipt of any Net Sales for the Product in the Territory, Kiniksa will [***] for such Quarter no later than [***] after its receipt of the Quarterly Expense Report from Regeneron (but shall not be obligated to make such payment earlier than [***] after the conclusion of such Quarter for the first three (3) Quarters of a Contract Year, or [***] following the end of the fourth (4^{th}) Quarter of each Contract Year). After receipt of the first Net Sales for the Product in the Territory, Kiniksa will deduct all Other Shared Expenses (including those incurred by Regeneron and reimbursed by Kiniksa) from Net Sales in its calculation of Profit. Kiniksa shall pay to Regeneron the amounts due as Regeneron Profit Split for such Quarter (such payment, the "Regeneron Profit Split Payment") [***] after its delivery of the Profit Payment Report for such Quarter (but shall not be obligated to make such payment earlier than [***] after such Quarter). If for any Quarter there is no Regeneron Profit Split Payment because there

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is no Profit, then Kiniksa will still reimburse Regeneron for the Other Shared Expenses incurred by Regeneron in such Quarter in accordance with this Section 9.4.5(c) (Payment to Regeneron).

(d) Payment to Regeneron of the Regeneron Third Party Proceeds Payment. No later than [***] after its delivery of the Third Party Proceeds Payment Report, Kiniksa shall pay to Regeneron the amounts due as the Regeneron Third Party Proceeds Split for such Quarter (such payment, the "Regeneron Third Party Proceeds Split Payment") but Kiniksa shall not obligated to make such payment earlier than [***] after such Quarter.

(e) No Offset. No expenses included or losses reflected in the Profit Payment Report in a Quarter shall be offset against Third Party Proceeds.

(f) Payments of Certain Invoiced Amounts. The Party obligated to make payments pursuant to Section 9.4.2 (Reimbursement of Manufacturing Costs), Section 9.4.3 (Kiniksa Incurred Expenses under Section 8.8.3 (Cost of Product Changes)), and Section 9.5.2 (Certain Commercial Shared Commercial Expenses [***], in each case, shall reimburse the other Party for the expenses no later than [***] after its receipt of an invoice or report delivered in accordance with Section 9.4.2 (Reimbursement of Manufacturing Costs), Section 9.4.3 (Kiniksa Incurred Expenses Under Section 8.8.3 (Cost of Product Changes), or Section 9.5.2 (Certain Commercial Shared Commercial Expenses [***], but shall not be obligated to make such payment earlier than [***] days after such Quarter.

9.4.6 **Invoices and Documentation**. The Parties shall agree upon the form of any necessary documentation relating to the Profit Split Arrangement hereunder so as to afford the Parties appropriate accounting treatment in relation to any of the transactions or payments contemplated hereunder.

9.4.7 **No Carryforward and Exception**.

(a) No Carryforward. If the Profits for any Quarter are negative, such that Kiniksa incurs a loss, then no such losses or components of expenses included in Profits in one Quarter shall be carried-forward an applied against Profits or the Regeneron Profit Split due in any subsequent Quarter.

(b) [***]. Notwithstanding Section 9.4.7(a) (No Carryforward), any [***] the Regeneron Profit Split in a Quarter because it would cause the Profits for such Quarter to be negative (such amount, the "Manufacturing Technology Transfer Cost Balance"), may be [***]; *provided that* Profits otherwise due in any subsequent Quarter may be only [***] to the extent

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[***] of the Profits and the Regeneron Profit Split that would have applied prior to the application of any of the [***].

9.4.8 [***] **Third Party License Payments** [***]. If [***], then [***]; *provided that* [***] as of the Effective Date. If [***], then [***]. In addition, [***], then [***] as of the Effective Date.

9.4.9 [***] **Expenses**. [***] Expenses may be included in the calculation of Profits in any Quarter.

9.5 [***] **Inclusion of Certain Shared Commercial Expenses.**

9.5.1 [***] **Inclusion**. For purposes of calculating the Regeneron Profit Split pursuant to Section 9.4 (Sharing of Profits), the aggregate of costs and expenses included under Section 1.189(b) and Section 1.189(f) of the definition of Shared Commercial Expenses for any Contract Year:

(a) [***];

(b) [***];

(c) [***]; and

(d) [***].

9.5.2 **Certain Shared Commercial Expenses** [***].

(a) Projected Net Sales. On an annual basis no later than September 30 of each Contract Year for which a [***] is expected to apply, Kiniksa shall prepare a good faith projected Net Sales for the Product in the Territory for the upcoming Contract Year ("Projected Net Sales") and reasonable supporting documentation underlying such projections and provide such projection and documentation to the JSC for its review and discussion.

(b) Calculation. For purposes of calculating the Certain Shared Commercial Expenses [***], the Projected Net Sales shall be used to calculate the applicable the Certain Shared Commercial Expenses [***] in the Quarter in accordance with the [***] set forth in Section 9.5.1 [***] Inclusion).

(c) Reports of Actuals. Within [***] following the end of each Contract Year in which a Certain Shared Commercial Expenses [***], Kiniksa shall deliver electronically to Regeneron a written report setting forth the actual Net Sales for the Product in the prior Contract Year, the actual Certain Commercial Shared Commercial Expenses [***] in the prior Contract Year (based on actual Net Sales during such Contract Year), and [***], for each Quarter in the previous Contract Year. Any

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adjustments in the Profit Split Arrangement [***] shall be made in accordance with Section 9.4.5(f) (Funds Flow).

9.5.3 **Exclusion** [***].

(a) Subsequent Indication [***]. In advance of each expected Marketing Approval for each subsequent Indication of a Product in the Kiniksa Field in the Territory after the receipt of U.S. Marketing Approval, Kiniksa shall prepare a [***] Commercial Plan for the launch and Commercialization of such Product in such Indication (a "Subsequent Indication Launch Plan") and submit such plan to the JSC for its review and discussion pursuant to Section 4.1.3(p) [***] and submit [***] to the JSC for its review and approval pursuant to Section 4.1.3(q). The JSC shall review consider in good faith for approval, each Subsequent Indication [***], and such approval by the JSC shall not be unreasonably withheld, conditioned, or delayed. If the JSC approves a Subsequent Indication [***] so approved. For clarity, if the JSC does not approve a Subsequent Indication [***] then Kiniksa may proceed [***].

(b) Expert Determination. If, following escalation to the Party's Executive Officers in accordance with Section 4.2.2(b) (Escalation), such Executive Officers cannot reach agreement on the appropriate Subsequent Indication [***], then either Party may submit the matter for resolution by [***] will select one or the other of the Party's proposed Subsequent Indication [***].

9.6 **Adjustments to FTE Rates**. Notwithstanding anything herein to the contrary, upon the reasonable request of either Party, the Parties shall meet to review the accuracy of any applicable FTE Rate in any country or Region. The Parties will share reasonable supporting documents and materials in connection with an assessment of such FTE Rate and to determine in good faith whether to adjust such FTE Rate in such country or Region.

9.7 **Late Payments**. All undisputed late payments made under this Agreement (including payments made pursuant to Section 9.1 (Upfront Payment), Section 9.2 (Development Milestones), Section 9.3 (Sharing of Third Party Proceeds), and Section 9.4 (Sharing of Profits)), shall earn annual interest, to the extent permitted by Applicable Law, from the date due until paid at a rate equal to [***], as quoted on [***] (or any other source agreed to by the Parties) effective for the date on which the payment was due, plus [***] percentage points (accrued monthly), unless such payments are disputed in good faith pursuant to Section 9.9 (Resolution of Payment Disputes).

9.8 **Taxes**. Any milestones, payments, or other amounts that are due or payable by one Party to the other Party pursuant to this Agreement ("Payments") shall not be reduced on the account of any taxes unless required by Applicable Law. The Party receiving a Payment (the "Payee Party") alone shall be responsible for paying any and all taxes (other than

withholding taxes required by Applicable Law to be paid by the Party making such Payment) (the "Paying Party") levied on account of, or measured in whole or in part by reference to, any Payments it receives. The Paying Party shall deduct or withhold from the Payments any taxes that it is required by Applicable Law to pay to any taxing authority. In such case, the Paying Party will promptly provide the Payee Party with copies of receipts or other evidence reasonably required and sufficient to allow the Payee Party to document such tax withholdings adequately for purposes of claiming foreign tax credits and similar benefits. The Parties will cooperate reasonably in completing and filing documents required under the provisions of any applicable tax laws or under any other applicable Law, in connection with the making of any required tax payment or withholding payment, or in connection with any claim to a refund of or credit for any such payment. The Parties will cooperate to minimize such taxes in accordance with Applicable Law, including using reasonable efforts to access the benefits of any applicable treaties. Such cooperation may include the Paying Party making payments from a single source in the U.S., where possible. . Apart from any withholding permitted under this Section 9.8 (Taxes) and those deductions expressly included in the definition of Net Sales, the amounts payable hereunder will not be reduced on account of any taxes, charges, duties, or other levies.

9.9 **Resolution of Payment Disputes**. In the event there is a dispute relating to any payment obligations or reports under this ARTICLE 9 (Payments), then the Party with the dispute shall have its representative on the JSC provide the other Party's representative on the JSC with written notice setting forth in reasonable detail the nature and factual basis for such good faith dispute and the Parties, through the JSC pursuant to Section 4.1.3(r) (Duties of the JSC), will seek to resolve the dispute as promptly as possible, but no later than [***] after such written notice is received. In the event that no resolution is reached by the JSC, then such matter will be shall be resolved in accordance with Section 17.1 (Disputes). The Parties agree that if there is a dispute regarding any payment amount due under this Agreement, then only the disputed amount shall be withheld from the payment and the paying Party shall pay the undisputed amount within the timeframes set forth in this ARTICLE 9 (Payments).

ARTICLE 10
INTELLECTUAL PROPERTY

10.1 **Ownership of Newly Created Intellectual Property**. The determination of whether Know-How or other inventions are invented by a Party for the purpose of allocating proprietary rights therein, shall, for purposes of this Agreement, be made in accordance with U.S. patent law or other Applicable Law in the United States irrespective of where such conception, discovery, development, or making occurs. As between the Parties, each Party will own any and all inventions, improvements, works, and Know-How invented, discovered, conceived, created, or otherwise generated (including as necessary to establish authorship (in case of publication and other copyrightable work), inventorship (in case of inventions, whether patentable or not) or ownership), solely by or on behalf of such Party or its Affiliates (or its or their respective directors, officers, employees, or agents), in the course of performing activities under this Agreement, and any and all Patent Rights and other Intellectual Property Rights thereto, and the Parties shall jointly own an equal and undivided interest in all Joint IP, [***], Kiniksa shall [***].

10.2 **Assignment and Covenants in Support of Assignment**. To the extent that any right, title, or interest in or to any Intellectual Property Right invented, discovered, conceived, created, reduced to practice, or otherwise generated under this Agreement vests in a Party or its Affiliate, by operation of law or otherwise, in a manner contrary to the agreed upon ownership as set forth in Section 10.1 (Ownership of Newly Created Intellectual Property), such Party (or its Affiliate) shall, and hereby does, irrevocably assign to the other Party any and all such rights, title, and interests in and to such Intellectual Property Right to the other Party without the need for any further action by any Party. In furtherance of the foregoing, each Party shall, upon request of the other, promptly undertake and perform (or cause its Affiliates and its and their respective employees or agents to promptly undertake and perform) such further actions as are reasonably necessary for Regeneron and Kiniksa, as between the Parties, to each perfect its title in any such Intellectual Property Right as set forth in Section 10.1 (Ownership of Newly Created Intellectual Property), including by causing the execution of any assignments or other legal documentation, or providing the other Party or its patent counsel with reasonable access to any employees or agents who may be inventors of such Intellectual Property Rights.

10.3 **Exploitation of Joint IP**. Subject to the other applicable provisions of this Agreement (including Section 2.1 (Regeneron Licenses to Kiniksa) and Section 2.2 (Kiniksa Licenses to Regeneron)), each Party shall otherwise enjoy an equal undivided right to exploit any and all Joint IP, including the right to use, practice, and otherwise exploit for research, development, manufacturing, commercial, and other purposes (including to grant licenses or other similar rights under) the Joint IP, without the need to seek consent from or account to the other Party (and, for clarity, neither Party shall be required to obtain the consent of the other Party with respect to the exploitation thereof anywhere in the world and, to the extent that such consent is required in any country in the world, such consent is hereby granted).

10.4 **Invention Disclosures; Updates and Exchange of Information**. Each Party shall promptly disclose to the other Party all Know-How and other Intellectual Property Rights that are invented, discovered, conceived, created, reduced to practice, or otherwise generated by such Party, its Affiliates, or their employees, agents, and consultants pursuant to this Agreement that is Product IP, Kiniksa Platform IP, or Joint IP. Each Party will share with the JSC in a timely manner any material additions to the Regeneron IP (including Product IP) and Kiniksa Product Data (including all clinical data) that are in a Party's possession and Control, but excluding Regeneron Manufacturing IP.

10.5 **Assignment Obligation**. All of the employees, officers, and consultants of each Party that are engaged in the performance of its obligations or exercise of its rights under this Agreement shall have executed written agreements (a) assigning to such Party ownership of all inventions and Intellectual Property Rights made during the course of or as the result of employment or provision of services, as applicable, (b) obligating the individual upon request to sign any documents to confirm or perfect such assignment and to cooperate in the preparation and prosecution of any Patent Applications disclosing or claiming such inventions, and (c) obligating the individual to obligations of confidentiality and non-use regarding Confidential Information that are at least as stringent as those undertaken by the Parties pursuant to ARTICLE 13 (Confidentiality).

10.6 **No Other Rights or Licenses**. The Parties agree that nothing in this Agreement, and no use by a Party of the other Party's Intellectual Property Rights pursuant to this Agreement, shall vest in a Party any right, title, or interest in or to the other Party's Intellectual Property Rights, other than the license rights expressly granted hereunder and the assignments expressly made hereunder.

10.7 **Prosecution and Maintenance of Patent Rights.**

10.7.1 **Regeneron Patent Rights and Joint Patent Rights**. During the Term, the Responsible Party shall be solely responsible for the filing, prosecution, maintenance or defense of the Regeneron Patent Rights and Joint Patent Rights in the Territory, using reasonable efforts to prosecute all Patent Applications forming part of Regeneron Patent Rights or Joint Patent Rights (as applicable) to grant with valid claims. The Responsible Party shall keep the Non-Responsible Party informed of all material developments in relation to Regeneron Patent Rights and Joint Patent Rights in the Territory and shall, provide the Non-Responsible Party with copies of relevant documents related to the filing, prosecution, and maintenance of Regeneron Patent Rights and Joint Patent Rights. The Responsible Party shall consult with the Non-Responsible Party and shall consider in good faith any reasonable recommendation made by the Non-Responsible Party in relation to the scope of filing and the prosecution of Regeneron Patent Rights and Joint Patent Rights, including, when making any submission to a patent office or otherwise in the conduct of any proceedings in relation to such Patent Rights.

10.7.2 **Existing Regeneron Patent Rights; Costs**. Prior to the date of receipt of U.S. Marketing Approval, and so long as is it the Responsible Party, Regeneron shall solely bear its Fully-Burdened Cost incurred in the course of prosecuting and maintaining the Regeneron Patent Rights Controlled by Regeneron or its Affiliates as of the Effective Date in the U.S. and Japan and all Patent Rights claiming priority thereto (the "Existing Regeneron Patent Rights"). Prior to the date of receipt of U.S. Marketing Approval, all of Regeneron's Out-of-Pocket Costs incurred in the course of prosecuting and maintaining the Regeneron Patent Rights, other than the Existing Regeneron Patent Rights and the Joint Patent Rights, in each case, shall be considered Other Shared Expenses, and after the date of receipt of U.S. Marketing Approval, all of either Party's Out-of-Pocket Costs incurred in the course of prosecuting and maintaining the Regeneron Patent Rights (including as the Non-Responsible Party) shall be considered Other Shared Expenses.

10.7.3 **Regeneron Patent Rights and Joint Patent Rights; Second Right**. Subject to Section 10.7.1 (Regeneron Patent Rights and Joint Patent Rights), in the event that the Responsible Party wishes to decline to file or, having filed, wishes to abandon, allow claims to issue in a Patent without reserving the right to file a continuing or divisional Patent Application, maintain or defend any pending Regeneron Patent Rights or Joint Patent Rights in the Territory, then the Responsible Party shall provide the Non-Responsible Party with written notice thereof. In the case where the Responsible Party has filed but is declining to further prosecute or maintain such Regeneron Patent Rights or Joint Patent Rights, such notice shall be given at least [***] days prior to the expiration of any official substantive deadline relating to such activities. Subject to Section 10.7.1 (Regeneron Patent Rights and Joint Patent Rights), in any of such circumstances the Non-Responsible Party shall have the right to decide that the Non-Responsible Party should file, prosecute and maintain such Regeneron Patent Rights in Regeneron's name or Joint Patent Rights in the names of both Parties and in such case the Non-Responsible Party shall give written notice to the Responsible Party. Upon receipt of any such notice from the Non-Responsible Party, the Responsible Party shall transfer to the Non-Responsible Party all its files relating to the relevant Regeneron Patent Rights or Joint Patent Right and execute any documents to otherwise necessary to transfer control of such filing, prosecution and maintenance to the Non-Responsible Party. Thereafter, the Non-Responsible Party shall be responsible for prosecuting and maintaining such Regeneron Patent Rights or Joint Patent Rights and shall be the "Responsible Party" for the purposes of this Section 10.7 (Prosecution and Maintenance of Patent Rights). In any case where the newly designated Responsible Party so files, prosecutes or maintains such Regeneron Patent Rights or Joint Patent Rights, the newly designated Non-Responsible Party shall provide all necessary cooperation and assistance to newly designated Responsible Party in relation to any such proceeding. All Out-of-Pocket Costs incurred by the newly designated Responsible Party pursuant to this Section

10.7.3 (Regeneron Patent Rights and Joint Patent Rights; Second Right) and the newly designated Non-Responsible Party in connection with transitioning activities to the newly designated Responsible Party pursuant to this Section 10.7.3 (Regeneron Patent Rights and Joint Patent Rights; Second Right) shall each be considered Other Shared Expenses.

10.7.4 **Qualifications**. The Parties acknowledge and agree that the rights and obligations of the Parties with respect to the planned Patent Application(s) set forth on Schedule 10.7.4, shall be subject to the additional conditions set forth therein. In the event of a conflict between Section 10.7.1 (Regeneron Patent Rights and Joint Patent Rights), Section 10.7.2 (Existing Regeneron Patent Rights; Costs) and Section 10.7.3 (Regeneron Patent Rights and Joint Patent Rights; Second Right) and Schedule 10.7.4, Schedule 10.7.4 shall control solely with respect to such planned Patent Application(s) and any resulting Patents.

10.7.5 **Neovii** [***].

(a) Kiniksa acknowledges that to the extent the Regeneron Patent Rights are necessary or useful to the Product in the Retained Field, such Patent Rights have been licensed to Neovii outside of the U.S. and Japan and Kiniksa's prosecution and maintenance rights in Section 10.7.1 (Regeneron Patent Rights and Joint Patent Rights) outside of the U.S. and Japan [***].

(b) Kiniksa acknowledges that to the extent the Regeneron Patent Rights are necessary or useful to the Product in the Retained Field, such Patent Rights have been licensed to Neovii outside of the U.S. and Japan [***], *provided that* [***].

10.7.6 **CREATE Act**. Neither Party shall have the right, without the prior written consent of the other Party, to invoke the Cooperative Research and Technology Enhancement Act of 2004, 35 U.S.C.103(c)(2)-(c)(3) (the "CREATE Act") with respect to any invention that is developed pursuant to this Agreement.

10.8 **Third Party Infringement Suits**. In the case where either Party believes that an infringement by a Third Party of Regeneron Patent Rights, Joint Patent Rights, or Patent Rights within the Kiniksa Platform IP may be occurring in one or more countries within the Territory, such Party shall disclose full details of the potential infringement to the other Party, subject to any necessary agreements to preserve the privileged nature of such communications.

10.8.1 **First Right**. The Responsible Party shall have the first right, but not the obligation, to conduct any such infringement claim and any proceedings including any counterclaim for invalidity or unenforceability or any declaratory judgment action and including the right to settle in its discretion; *provided that* the Responsible Party shall consult with the Non-Responsible Party and consider

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in good faith any reasonable comments made by the Non-Responsible Party in relation to the conduct of such proceedings, and *provided, further*, that such settlement does not include a license under Regeneron Patent Rights or Joint Patent Rights and does include any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or a Joint Patent Rights or bind the Non-Responsible Party to any monetary settlements. If the settlement includes a license under Regeneron Patent Rights or Joint Patent Rights or any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or Joint Patent Right, then the Non-Responsible Party's consent to the terms of such license or any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or a Joint Patent Rights shall be required, such consent not to be unreasonably withheld, conditioned or delayed. Where the Responsible Party decides to commence proceedings in relation to Regeneron Patent Rights or Joint Patent Rights, it shall be entitled to require the Non-Responsible Party to join the Responsible Party as co-plaintiff. In such case the Non-Responsible Party shall provide all necessary assistance to the Responsible Party in relation to any such proceeding.

10.8.2 **Second Right**. If the Responsible Party fails to undertake any such proceedings, then the Non-Responsible Party may give the Responsible Party notice and reasonable supporting evidence of infringement by a Third Party requesting the Responsible Party to undertake such proceedings within ninety (90) days of the date of notice or a shorter period if required to comply with Applicable Law, and if the Responsible Party decides not to do so, then with respect to infringement of any Regeneron Patent Rights or Joint Patent Right, the Non-Responsible Party shall be entitled to do so at its own cost and expense, subject to Section 10.8.6 (Reimbursement by the Responsible Party). Thereafter, in such case, the Non-Responsible Party shall be responsible for enforcing Regeneron Patent Rights or Joint Patent Rights and shall be the "Responsible Party" for the purposes of this Section 10.8.2 (Second Right). The newly designated Responsible Party shall control the conduct of any claim or proceedings including any counterclaim for invalidity or unenforceability or any declaratory judgment action, *provided*, *however* that such newly designated Responsible Party shall consult with and consider in good faith any reasonable comments made by the newly designated Non-Responsible Party in relation to the conduct of such proceedings. The Non-Responsible Party shall provide all necessary assistance to the Responsible Party in relation to such proceedings. The Responsible Party shall have sole right to settle such proceedings including any counterclaim for invalidity or unenforceability; *provided that* such settlement does not include a license under Regeneron Patent Rights or Joint Patent Rights and does not include any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or a Joint Patent Rights or bind the Non-Responsible Party to any monetary settlements. If the settlement includes a license under Regeneron Patent Rights or Joint Patent Rights or any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or

Joint Patent Right, then the Non-Responsible Party's consent to the terms of such license or any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or a Joint Patent Rights shall be required, such consent not to be unreasonably withheld, conditioned or delayed.

10.8.3 **Kiniksa Platform IP**. For clarity, Regeneron shall not have rights under Section 10.8.1 (First Right) or Section 10.8.2 (Second Right) to enforce the Kiniksa Platform IP.

10.8.4 **Neovii** [***].

(a) Notwithstanding Section 10.8.1 (First Right) or Section 10.8.2 (Second Right), Kiniksa shall not have any rights to enforce the Regeneron Patent Rights as the Responsible Party if the infringement of the Regeneron Patent Rights is solely in the Retained Field, solely in the Retained Territory or solely in the Regeneron Retained EEO Field.

(b) Kiniksa acknowledges that to the extent the Regeneron Patent Rights are necessary or useful to the Product in the Retained Field, such Patent Rights have been licensed to Neovii, [***].

(c) Kiniksa acknowledges that to the extent the Regeneron Patent Rights are necessary or useful to the Product in the Retained Field, such Patent Rights have been licensed to Neovii [***], *provided that* [***].

10.8.5 **Allocation of Recoveries**. If a Party succeeds in any such infringement proceedings enforcing Regeneron Patent Rights or Joint Patent Rights, whether at trial or by way of settlement, such Party shall be entitled to retain such part of any award of costs and damages made in such proceedings or settlement sum as is equal to such Party's costs of taking the proceedings and the such Party shall be entitled to retain the balance received by the such Party less an amount equivalent to the amount that would have been due to the other Party on the balance as if they were an amount to be distributed under the Profit Split which amount shall be paid to the other Party. Notwithstanding the foregoing [***], *provided, however* [***].

10.8.6 **Reimbursement by the Responsible Party**. All Out-of-Pocket Costs incurred by the Responsible Party pursuant to Section 10.8.1 (First Right) or Section 10.8.2 (Second Right) shall be considered Other Shared Expenses, but only to the extent that (i) [***].

10.9 **Regeneron Manufacturing Patents Rights Not Specific to a Product.** Notwithstanding Section 10.7.1 (Regeneron Patent Rights and Joint Patent Rights), Section 10.7.3 (Regeneron Patent Rights and Joint Patent Rights; Second Right), Section 10.8.1 (First Right), and Section 10.8.2 (Second Right), Kiniksa shall not have any Patent prosecution and maintenance rights or Patent enforcement rights to the extent that a Regeneron

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.

Manufacturing Patent Right relates to methods of Manufacturing that are not specific to the Product and that have applicability to other products. Notwithstanding Section 10.7.1 (Regeneron Patent Rights and Joint Patent Rights) and Section 10.7.3 (Regeneron Patent Rights and Joint Patent Rights; Second Right), Kiniksa shall not be obligated to reimburse Regeneron for Regeneron's Out-of-Pocket Costs incurred in the course of prosecuting and maintaining the Regeneron Manufacturing Patent Rights covered by this Section 10.9 (Regeneron Manufacturing Patent Rights Not Specific to a Product) or enforcing or participating in the recovery of the Regeneron Manufacturing Patent Rights covered by this Section 10.9 (Regeneron Manufacturing Patent Rights Not Specific to a Product).

10.10 **Patent Marking.** Each Party shall comply with the patent marking statutes in each country in which a Product is made, offered for sale, sold, or imported by such Party, its Affiliates or licensees or sublicensees.

10.11 **Third Party Claims**.

10.11.1 **Notice of Infringement Claims**.

(a) If either Party or its Affiliates learns of an allegation that the Development or Commercialization of the Product in the Kiniksa Field or the Manufacturing of the Product, in each case, in the Territory, under this Agreement infringes or otherwise violates the Intellectual Property Rights of any Third Party in the Territory ("Infringement Claim"), then such Party must promptly notify the other Party in writing of such Infringement Claim. As soon as reasonably practicable after the receipt of such notice and at all times thereafter, the Parties will meet and consider the appropriate course of action with respect to such Infringement Claim. In any such instance, each Party will have the right to defend any action naming it; *provided, however*, the Parties will at all times cooperate, share all material notices and filings in a timely manner, provide all reasonable assistance to each other and use Commercially Reasonable Efforts to agree upon an appropriate course of action, including, as appropriate, the preparation of material court filings and any discussions concerning a potential defense or settlement of any such Infringement Claim. The rights and obligations in this Section 10.11 (Third Party Claims) will apply even if only one Party defends any Infringement

Claims commenced by a Third Party in the Territory claiming that the Development, Manufacture, or Commercialization of a Product under this Agreement infringes or otherwise violates any Intellectual Property Rights of any Third Party.

(b) Notwithstanding Section 10.11.1(a) (Notice of Infringement Claims), any settlement by a Party of an Infringement Claim that includes a license under Regeneron Patent Rights or Joint Patent Rights or any admission concerning claim invalidity or unenforceability of a

Regeneron Patent Right or Joint Patent Right, requires the consent of the non-settling Party to the terms of such license or any admission concerning claim invalidity or unenforceability of a Regeneron Patent Right or a Joint Patent Rights, such consent not to be unreasonably withheld, conditioned or delayed. Additionally, in no event may the settling Party bind the non-settling Party to any monetary settlement without the consent of the non-settling Party.

10.11.2 **Coordination with Neovii**. Kiniksa acknowledges that an Infringement Claim may also name Neovii or otherwise triggers certain rights of Neovii under the Neovii License Agreement (which rights are equivalent to the ones set forth in Section 10.11.1(a) (Notice of Infringement Claims)). In such case, Regeneron shall use Commercially Reasonable Efforts to coordinate between Kiniksa and Neovii to ensure that Kiniksa's rights under Section 10.11.1(a) (Notice of Infringement Claims) are preserved.

10.11.3 **Reimbursement** [***]. Out-of-Pocket Costs incurred by either Party in connection with the defense of an Infringement Claim (including any nullification, declaratory judgment, revocation, or opposition proceeding against any such Patents or other rights in response to prospective or actual Infringement Claim) shall [***].

10.12 **Compliance with Third Party Licenses**. The Parties acknowledge and agree that Regeneron does not have the right to (a) prosecute and maintain any Patent Rights licensed to it under the Existing Licenses, or (b) enforce Patent Rights and Know-How licensed to it under the Existing Licenses against Third Party infringement, and therefore the rights granted by Regeneron to Kiniksa under this ARTICLE 10 (Intellectual Property) are subject to this Section 10.12 (Compliance with Third Party Licenses).

10.13 **Product Trademarks**.

10.13.1 **Kiniksa's Use of Product Trademarks**. At Kiniksa's sole discretion (but subject to the following sentence), Kiniksa may use the Existing Product Trademarks or New Product Trademarks in connection with the Development and Commercialization of the Product in the Kiniksa Field throughout the Territory (if necessary in the script of a different language). Notwithstanding the foregoing, [***] (a) [***], or (b) [***]. Regeneron shall coordinate the use of Product Trademarks [***] to ensure there is no confusion [***]. Upon Kiniksa's request reasonably in advance of its filling and finishing of Product for Commercialization, Regeneron shall provide Kiniksa with the identity of the Existing Product Trademarks and the countries or Regions referred to in clause (a) of the second sentence of this Section 10.13.1 (Kiniksa's Use Product Trademarks).

10.13.2 **Ownership of Existing Product Trademarks**. Regeneron (or its local Affiliates, as appropriate) will own and retain all rights, title, and interests in and

to the Existing Product Trademarks, together with all associated domain names and all goodwill related thereto. At Kiniksa's request, Regeneron shall use reasonable efforts to take all actions necessary under Applicable Laws to register Kiniksa as a licensee of the Existing Product Trademarks in a country or Region in the Territory, and Kiniksa shall reasonably cooperate with Regeneron in furtherance thereof. If, as a result of the requirement of any local regulation or Regulatory Authority in a particular country or Region in the Territory, Kiniksa must own the Existing Product Trademarks for such country or Region, then Regeneron shall assign the rights to the Existing Product Trademarks in such country or Region to satisfy the requirement or regulation. If Regeneron so assigns ownership of any Existing Product Trademark to Kiniksa in a country or Region in the Territory, and Kiniksa does not Commercialize the Product under, or otherwise use, such Existing Product Trademark in such country or Region for a period of at least [***] years, then, upon Regeneron's written request, Kiniksa will assign to Regeneron its rights, title, and interests in and to such Existing Product Trademark in such country or Region.

10.13.3 **Ownership, Prosecution, and Maintenance of Existing Product Trademarks**. Regeneron will be responsible for prosecuting and maintaining the Existing Product Trademarks in each country or Region in the Territory in which Kiniksa intends to Commercialize a Product (as communicated in writing to Regeneron in good faith) using reasonable efforts. Kiniksa will, to the extent required, provide such assistance and execute such documents in any such country or Region in the Territory as reasonably requested by Regeneron to allow Regeneron to file, register, or maintain such Existing Product Trademarks in respect of the Product in the Kiniksa Field in such country or Region in the Territory. Regeneron's Out-of-Pocket Costs incurred in the course of prosecuting and maintaining such Existing Product

Trademarks in such countries or Regions shall be considered Other Shared Expenses. Regeneron shall keep Kiniksa advised of all activity related to such filings, registrations, maintenance, and prosecution of such Existing Product Trademarks in such countries or Regions, and shall coordinate such activity with Kiniksa.

10.13.4 **Kiniksa Step-In; Costs**. In the event that Regeneron elects not to file any Existing Product Trademark in a country or Region in the Territory in which Kiniksa intends to Commercialize a Product (as communicated in writing to Regeneron in good faith), or elects not to prosecute or maintain any Existing Product Trademark filed in any such country or Region in the Territory, then, in each case, Regeneron shall provide reasonable prior written notice to Kiniksa of its intention not to file, prosecute, or maintain any such Product Trademark in such country or Region, and Kiniksa shall have the right to do so with respect to such Product in the Kiniksa Field on behalf of Regeneron, subject to consultation and cooperation with Regeneron. Kiniksa's Out-of-Pocket Costs incurred pursuant to this Section 10.13.4 (Kiniksa Step-In; Costs) shall be considered Other Shared Expenses.

10.13.5 **New Product Trademarks**. Kiniksa (or its local Affiliate, as appropriate) will own and retain all rights, title, and interests in and to the New Product Trademarks, together with all associated domain names and all goodwill related thereto. Kiniksa will be responsible for prosecuting and maintaining the New Product Trademarks in each country or Region in the Territory using reasonable efforts. Regeneron will, to the extent required, provide such assistance and execute such documents in any such country or Region in the Territory as reasonably requested by Kiniksa to allow Kiniksa to file, register, or maintain such New Product Trademarks in respect of the Product in the Kiniksa Field in such country or Region. Kiniksa's Out-of-Pocket Costs incurred pursuant to this Section 10.13.5 (New Product Trademarks) shall be considered Other Shared Expenses.

10.13.6 **Use of Trademarks**. Each Party agrees that the use of the other Party's Trademarks shall comply with all Applicable Law and such other Party's Trademark policies. Each Party will refrain from any use of the other's Trademarks in a manner that threatens to damage the goodwill associated with the respective Trademarks or that threatens to tarnish the reputation or otherwise reflect unfavorably upon the owner of the Trademarks. Neither Party shall, during the Term, anywhere in the world, take any action that in the other Party's sole and absolute discretion impairs or contests or is likely to impair or contest the validity of the other Party's rights, title, or interests in and to its Trademarks, including using, or filing an application to register, any word, mark, symbol or device, or any combination thereof, that is confusingly similar to or dilutes the distinctiveness of any of the other Party's Trademarks.

10.13.7 **Quality Control; Inspection**. Kiniksa shall maintain standards regarding the nature and quality of the Product in accordance with such quality control standards and procedures provided by Regeneron during the Term. [***]

10.13.8 **Neovii** [***]. Kiniksa acknowledges that [***], *provided that* [***].

10.13.9 **Product Trademark Domain Names**. Except as required as set forth in this Section 10.13.1 (Ownership of Existing Product Trademarks), Kiniksa and its Affiliates shall not register any domain names consisting of or containing any Existing Product Trademark or any other trademark owned by Regeneron. Kiniksa shall have the exclusive right to register the New Product Trademarks as domain names, which shall mean all internet domain names under all existing top level domains and Regeneron and its Affiliates shall not register any domain names consisting of or containing any New Product Trademark or any other trademark owned by Kiniksa.

10.13.10 **Product Notices**. All uses by Kiniksa of the Existing Product Trademarks owned by Regeneron shall be accompanied by a statement that the Existing Product Trademark is a registered trademark of Regeneron.

ARTICLE 11
BOOKS, RECORDS AND INSPECTIONS; AUDITS AND ADJUSTMENTS

11.1 **Books and Records**. Each Party shall keep complete and accurate books of record and account pertaining to the activities to be conducted and payments to be made under this Agreement ("Records") in which full, true, and correct entries (in conformity with GAAP) shall be made in sufficient detail to permit the other Party ("Auditing Party") to confirm the accuracy of all amounts payable or owed pursuant to this Agreement (including the utilization of FTEs, the determination of the Commercial Overhead Charge, COGS, Other Shared Expenses, and Shared Commercial Expenses, the allocation of personnel under this Agreement, and any royalties due to Kiniksa pursuant to Section 16.8.2(b) (Royalty Payments to Kiniksa)), and such Records shall be maintained (in such form as may be available) for a period of no less than [***] years following the end of the period to which they pertain. Each Party shall keep its Records in a readily available and organized form to allow an independent auditor to verify the accuracy of all financial, accounting and numerical information provided in an efficient manner. To the extent a Party subject to an audit ("Audited Party") is not in compliance with the foregoing, the Audited Party shall be responsible for any additional fees charged by the independent auditor to the Auditing Party as a result of additional time spent by the independent auditor assembling or organizing such information. Each Party

shall, and shall cause each of its respective Affiliates to, permit auditors to visit, inspect and examine, during regular business hours and under the guidance of officers of the Audited Party, the Records of such Party or such Affiliate, as provided in Section 11.2 (Audits and Adjustments).

11.2 **Audits and Adjustments**.

11.2.1 **Right to Audit**. Each Party shall have the right and at its own expense, upon no less than [***] days' advance written notice and at such reasonable times as the Party shall request, not more than once per Contract Year-period and not more than once during any Contract Year unless a material discrepancy is found in any such audit of such period, in which case additional audits of such period may be conducted until no material discrepancies are found, to have the Records of the other Party audited by an independent "Big Four" (or equivalent) accounting firm, reasonably acceptable to each Party to the extent relating to this Agreement for the preceding [***] years. The Auditing Party shall cause the designated accounting firm to enter into a written confidentiality agreement with the Audited Party containing confidentiality and non-use provisions substantially the same as those set forth herein, and to limit its audit of the Auditing Party solely to those Records pertaining to payments made, or activities conducted, this Agreement.

11.2.2 **Audit Results**. The accounting firm will first provide the results of any audit in writing to the Audited Party, and then will be further instructed to redact any proprietary information of the Audited Party not relevant to verifying the accuracy of the applicable reports prior to providing the audit report to the

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Auditing Party. Any audit shall be final and binding upon the Parties, unless disputed by a Party within [***] days after delivery of the applicable report. If a Party over billed or underpaid an amount due under this Agreement resulting in a cumulative discrepancy during any Contract Year of more than [***], then it shall also reimburse the other Party for the reasonable Out-of-Pocket Costs of such audit (with the cost of the audit to be paid by the Party initiating the audit in all other cases). Such accountants shall disclose to the Party requesting the audit a summary of its review and findings. The summary and findings, and all audit reports, in each case, shall be subject to the confidentiality provisions contained in ARTICLE 13 (Confidentiality).

11.2.3 **Overbilling; Underpayment**. If any examination or audit of the Records described above discloses an over billing or underpayment of amounts due hereunder, then unless the result of the audit is contested pursuant to Section 11.2.4 (Disputes), then the Party that overbilled or underpaid shall pay the same (plus interest thereon at the default interest rate set forth in Section 9.7 (Late Payments) from the date of such overbilling or underpayment through the date of payment of the amount required to be paid pursuant to this Section 11.2.3 (Overbilling; Underpayment)) to the Party entitled thereto within [***] days after receipt of the written results of such audit pursuant to this Section 11.2 (Audits and Adjustments).

11.2.4 **Disputes**. Any disputes with respect to the results of any audit conducted under Section 11.2 (Audits and Adjustments) shall be elevated to the JSC and resolved in accordance with Section 4.1.3(w) (Duties of the JSC).

11.3 **GAAP**. Except as otherwise provided herein, including Section 9.3 (Sharing of Third Party Proceeds), all costs and expenses and other financial determinations with respect to this Agreement shall be determined in accordance with GAAP, as generally and consistently applied by a Party throughout its organization.

ARTICLE 12
REPRESENTATIONS, WARRANTIES AND COVENANTS

12.1 **Joint Representations and Warranties**. Each Party represents and warrants to the other Party, as of the Effective Date, as follows: (a) it is duly organized and validly existing under the laws of its jurisdiction of incorporation; (b) it has full corporate power and authority and has taken all corporate action necessary to enter into and perform this Agreement; (c) the execution and performance by it of its obligations hereunder will not constitute a breach of, or conflict with, any Applicable Law, its organizational documents nor any other material agreement or arrangement, whether written or oral, by which it is bound or requirement of Applicable Laws; (d) this Agreement is its legal, valid, and binding obligation, enforceable in accordance with the terms and conditions hereof (subject to Applicable Laws of bankruptcy); and (e) no broker, finder or investment banker is entitled to any brokerage, finder's or other fee in connection with this Agreement or the transactions contemplated hereby.

12.2 **Knowledge of Pending or Threatened Litigation**. Each Party represents and warrants to the other Party that, as of the Effective Date, there is no announced investigation, suit, action or proceeding pending or, to such Party's Knowledge, threatened, against such Party before or by any court, arbitrator, or Governmental Authority that, individually or in the aggregate, is reasonably expected to (a) materially impair the ability of such Party to perform its obligations under this Agreement, or (b) prevent or materially delay or alter the consummation of any or all of the transactions contemplated hereby.

12.3 **No Conflict**. [***], each Party represents and warrants to the other Party that, as of the Effective Date, it has not entered into any agreement with any Third Party that is in material conflict with the licenses and other rights granted to the other Party under this Agreement, and has not taken any action that would in any way prevent it from granting the licenses and other rights granted to the other Party under this Agreement, or that would otherwise materially conflict with or materially adversely affect the licenses and other rights granted to the other Party under this Agreement, and its performance and execution of this Agreement will not result in a material breach of any other contract to which it is a party.

12.4 **Additional Regeneron Representations, Warranties, and Covenants of Regeneron**. Regeneron additionally represents and warrants to Kiniksa that, as of the Effective Date:

12.4.1 **Ownership or Control**. Except with respect to Patent Rights and Know-How licensed to Regeneron under the Existing Licenses, (a) Regeneron or its Affiliate Controls the Regeneron IP, and (b) Regeneron owns all right, title, and interest in and to all Regeneron Patent Rights in existence as of the Effective Date. Schedule 1.165 lists all Patent Rights Controlled by Regeneron or any of its Affiliates as of the Effective Date that qualify as Regeneron Patent Rights based on the definition thereof, [***];

12.4.2 **Sufficient Rights**. [***], Regeneron has sufficient legal or beneficial title, ownership, or license, free and clear from any mortgages, pledges, liens, security interests, encumbrances, charges, or claims of any kind, as necessary to grant the licenses to Kiniksa as contemplated by this Agreement;

12.4.3 **Existing Product Trademarks**. Schedule 1.58 includes all Trademarks Controlled by Regeneron and used in connection with the Commercialization of any Product as of the Effective Date;

12.4.4 **No Infringement Claims**. To Regeneron's Knowledge there is no pending litigation alleging, and Regeneron has not received written notice that alleges that any of Regeneron's making, using, selling, offering to sell, or importing of the Product has violated, or would violate, a Valid Claim of an issued and unexpired Patent Right of any Third Party in the Territory;

12.4.5 **No Misappropriation**. To Regeneron's knowledge, the Development, Manufacture, and Commercialization of the Product as of the Effective Date has

not constituted or involved the misappropriation of the trade secrets of a Third Party;

12.4.6 **Validity and Enforceability of Regeneron Patent Rights**. To Regeneron's Knowledge, the issued and unexpired Regeneron Patent Rights are not invalid or unenforceable, in whole or part;

12.4.7 **Prosecution and Maintenance**. The Regeneron Patent Rights have been prosecuted with the respective patent offices in accordance with Applicable Law, and to Regeneron's Knowledge, all fees necessary to maintain such Regeneron Patent Rights have been paid on or before the due date for such payment;

12.4.8 **Invention Assignments**. To Regeneron's Knowledge, each individual who is an inventor of, or otherwise contributed in a material manner to the creation or development of, any Regeneron Patent Rights has assigned to Regeneron all of his or her interest therein;

12.4.9 **No Challenges**. Regeneron has not received any written notice of (a) any threatened litigation seeking to invalidate or otherwise challenge any Regeneron Patent Rights or Regeneron's rights therein, or (b) any pending re-examination, opposition, interference, or litigation proceedings with respect to any Regeneron Patent Right;

12.4.10 **No Untrue Statements**. To Regeneron's Knowledge, neither Regeneron nor any officer, employee, or agent of Regeneron has knowingly made an untrue statement of a material fact to any Regulatory Authority in the Territory with respect to the Product or knowingly failed to disclose a material fact required to be disclosed to any Regulatory Authority in the Territory with respect to the Product, in each case, that could reasonably be expected to provide a basis for the FDA to invoke its policy respecting "Fraud, Untrue Statements of Material Facts, Bribery, and Illegal Gratuities", set forth in 56 Fed.Reg.46191 (September 10, 1991) and any amendments thereto or any analogous laws or policies in the Territory;

12.4.11 **Debarment**. (a) Regeneron has not been debarred or suspended under 21 U.S.C.§335(a) or (b), is not the subject of a conviction described in Section 306 of the FD&C Act, has not been and is not excluded from a federal or governmental health care program, debarred from federal contracting, convicted of or *pled nolo contendere* to any felony, or to any federal or state legal violation (including misdemeanors) relating to prescription drug products or fraud, is not subject to OFAC sanctions or on the OFAC list of specially designated nationals, and is not subject to any similar sanction of any Governmental Authority in the Territory ("Debarred"), and to Regeneron's Knowledge, no proceeding that could result in it being Debarred is pending, and neither it nor any of its Affiliates has used, in any capacity in the performance of obligations

relating to any Product, any employee, subcontractor, consultant, agent, representative, or other person who has been Debarred;

12.4.12 **No Bribery**. Neither Regeneron nor its employees and agents have, directly or indirectly through Third Parties, knowingly paid, promised or offered to pay, or authorized the payment of any money or given any promise or offer to give, or authorized the giving of anything of value, to a Public Official or Entity or other Person for purposes of corruptly obtaining or retaining business for or with, or directing business to, any Person by (a) influencing any official act, decision or omission of such Public Official or Entity; (b) inducing such Public Official or Entity to do or omit to do any act in violation of the lawful duty of such Public Official or Entity; (c) securing any improper advantage; or (d) inducing such Public Official or Entity to affect or influence any act or decision of another Public Official or Entity;

12.4.13 **No Kickbacks**. Neither Regeneron nor its employees and agents have knowingly promised, offered, or provided any corrupt payment, gratuity, emolument, bribe, kickback, excessive gift, or hospitality or other illegal or unethical benefit to a customer or a Third Party customer or to a Public Official or Entity;

12.4.14 **Existing Licenses**. Regeneron is not party to any agreement with any Third Party pursuant to which Regeneron has received a license under any Patents Rights or Know-How included in the Regeneron IP, [***]. Except with respect to the Existing Licenses, neither Regeneron nor any of its Affiliates is a party to any agreement that imposes a royalty or other similar contingent payments (*e.g.*, milestones) on the Development, Manufacture, or Commercialization of the Product (as it exists as of the Effective Date) in the Kiniksa Field or the Regeneron Field in the Territory; and

12.4.15 **No Breach of Existing Licenses**. Regeneron and its Affiliates have not received any written notice of material breach of any of the Existing Licenses. [***], to Regeneron's Knowledge there have been no acts or omissions by Regeneron or its Affiliates that constitute a material breach of an Existing License.

12.5 **Mutual Covenants**. Each Party hereby covenants to the other Party as follows:

12.5.1 it will not during the Term grant any right or license to any Third Party that would be inconsistent with or in conflict with or in derogation of the rights granted to the other Party under this Agreement, and will not take any action that would conflict with its obligations to the other Party under this Agreement;

12.5.2 neither Party will use the Patent Rights, Know-How, materials, or Confidential Information of the other Party outside the scope of the licenses and rights granted to it under this Agreement; and

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12.5.3 it will not engage, in any capacity in connection with this Agreement or any ancillary agreements, any officer, employee, contractor, consultant, agent, representative, or other person who has been Debarred. Each Party will inform the other Party in writing promptly if it or any person engaged by it or any of its Affiliates who is performing any obligations under this Agreement or any ancillary agreements is Debarred, or if any action, suit, claim, investigation, or legal or administrative proceeding is pending or, to each Party's Knowledge, is threatened, pursuant to which a Party, any of its Affiliates or any such person performing obligations hereunder or thereunder may become Debarred.

12.6 **Compliance with Laws**.

12.6.1 **Anti-Corruption**. Each Party agrees, in its performance of its obligations under this Agreement, to comply, and to cause its Affiliates to comply, with all Applicable Laws, including the FCPA, U.S. Export Control Laws, and Anti-Corruption Laws. Each Party shall not knowingly take any action that would cause the other Party to be in violation of the FCPA, U.S. Export Control Laws, or any other applicable Anti-Corruption Laws. Further, each Party shall immediately notify the other Party if such Party has any information or suspicion that there may be a violation of the FCPA or any other Anti-Corruption Law in connection with the performance of activities under this Agreement.

12.6.2 **No Bribery**. Each Party and its employees and agents shall not, directly or indirectly through Third Parties, knowingly pay, promise or offer to pay, or authorize the payment of, any money or give any promise or offer to give, or authorize the giving of anything of value, to a Public Official or Entity or other person for purposes of corruptly obtaining or retaining business for or with, or directing business to, any Person, including either Party, by (a) influencing any official act, decision or omission of such Public Official or Entity; (b) inducing such Public Official or Entity to do or omit to do any act in violation of the lawful duty of such Public Official or Entity; (c) securing any improper advantage; or (d) inducing such Public Official or Entity to affect or influence any act or decision of another Public Official or Entity.

12.6.3 **No Kickbacks**. Each Party and its employees and agents have not and shall not knowingly promise, offer or provide any corrupt payment, gratuity, emolument, bribe, kickback, excessive gift or hospitality or other illegal or unethical benefit to a customer or a Third Party customer or to a Public Official or Entity. In addition, each Party and its employees and agents shall ensure that no part of any payment, commission, reimbursement or fee paid by either Party pursuant to this Agreement or otherwise will be used knowingly as a corrupt payment, gratuity, emolument, bribe, kickback, excessive gift or hospitality or other illegal or unethical benefit to a customer or to Third Party customer or to a Public Official or Entity.

12.7 **Additional Covenants of Regeneron**.

12.7.1 **No Encumbrances of Regeneron IP**. Regeneron further covenants to Kiniksa as follows other than in connection with an assignment or other transfer of this Agreement as permitted under Section 17.9 (Assignment), Regeneron shall not assign, transfer or convey its rights to the Regeneron IP or otherwise encumber Kiniksa's license to the Regeneron IP that (in the case of Regeneron Patent Rights) Cover or (in the case of Know-How) is used in the Development or Commercialization of a Product in the Kiniksa Field in the Territory for as long as Kiniksa retains any license or other rights therein by virtue of this Agreement without the prior written consent of Kiniksa, such consent not to be unreasonably withheld, conditioned, or delayed.

12.7.2 **Maintenance of Rights Under Existing Licenses**. [***], Regeneron will not, and will cause its Affiliates not to, without Kiniksa's prior written consent, (a) commit any acts or permit the occurrence of any omissions that would cause breach or termination of any Existing License, or (b) amend or otherwise modify or permit to be amended or modified, any Existing License in a manner that would adversely affect the rights granted to Kiniksa under this Agreement.

12.8 **Disclaimer of Warranties**. EXCEPT AS OTHERWISE SPECIFICALLY PROVIDED IN THIS AGREEMENT OR THE SUPPLY AGREEMENTS, NEITHER PARTY MAKES ANY REPRESENTATIONS OR WARRANTIES, EXPRESS, IMPLIED, STATUTORY OR OTHERWISE, CONCERNING THE SUCCESS OR POTENTIAL SUCCESS OF THE DEVELOPMENT, COMMERCIALIZATION, MANUFACTURE, MARKETING, OR SALE OF ANY PRODUCT OR ANY INTELLECTUAL PROPERTY RIGHTS. EXCEPT AS EXPRESSLY SET FORTH HEREIN, EACH PARTY EXPRESSLY DISCLAIMS ANY AND ALL WARRANTIES OF ANY KIND, EXPRESS OR IMPLIED, INCLUDING THE WARRANTIES OF NON-INFRINGEMENT, MERCHANTABILITY, AND FITNESS FOR A PARTICULAR PURPOSE.

ARTICLE 13
CONFIDENTIALITY

13.1 **Confidential Information**. During the Term and for a period of [***] years thereafter, each Party and its Affiliates (in such capacity, collectively, the "Receiving Party") shall, and shall cause its officers, directors, employees, and agents to, keep confidential, and other than as provided herein, shall not publish or otherwise disclose, directly or indirectly, any confidential or proprietary information, including any scientific, clinical, regulatory, manufacturing, marketing, financial, and commercial information or data, controlled by the other Party or its Affiliates (in such capacity, collectively, the "Disclosing Party"), whether communicated in writing or orally or by any other method in tangible or intangible form, that is disclosed pursuant to this Agreement (the "Confidential Information"). Each Party and its Affiliates shall use the Confidential Information of the other Party and its Affiliates solely for the purpose of exercising its

rights and performing its obligations hereunder. For purposes of this Agreement, all confidential information disclosed by a Party under the terms of the Mutual Confidentiality Agreement between the Parties dated June 8, 2016, as amended on May 23, 2017 ("CDA") will be Confidential Information of such Party and treated as if disclosed hereunder and shall be subject to the terms of this Agreement. Each Party covenants that neither it nor any of its respective Affiliates shall disclose any Confidential Information of the other Party to any Third Party except (a) to its employees, agents, consultants, or any other Person under its authorization; provided that such employees, agents, consultants, or other Persons are subject in writing (or by explicit professional obligations such as the attorney-client relationship) to confidentiality obligations applicable to such Confidential Information substantially the same as those set forth herein, (b) as approved by both Parties hereunder in writing, (c) to investors, prospective investors, lenders, prospective lenders, financing sources, prospective financing sources, prospective acquirers, permitted sublicensees, prospective sublicensees, financial or legal advisors, or subcontractors; provided that such persons agree in writing (or by explicit professional obligations such as the attorney-client relationship) to confidentiality obligations applicable to such Confidential Information substantially the same as those set forth herein, and (d) as set forth elsewhere in this Agreement, including to subcontractors in accordance with Section 2.9 (Subcontracting). Each Party will ensure that such Party's Affiliates, investors, prospective investors, lenders, prospective lenders, acquirors, prospective acquirors, financing sources, prospective financing sources, permitted sublicensees, prospective sublicensees, employees, consultants, agents, consultants, and subcontractors comply with these obligations. Regeneron Know-How is the Confidential Information of Regeneron. Kiniksa Know-How is the Confidential Information of Kiniksa, and Joint Know-How and the terms of this Agreement is the Confidential Information of each Party. Joint Know-How shall be Confidential Information of each Party; provided that, except as expressly permitted herein, the Joint Know-How may be used by each Party as provided herein, but may not be disclosed to Third Parties without the prior written consent of the other Party. Notwithstanding the foregoing, without Regeneron's prior written consent (such consent not to be unreasonably withheld, conditioned, or delayed to the extent requested in connection with any Formulation Development Activities), Kiniksa shall not have the right to disclose Regeneron Manufacturing Know-How to any Third Party, except in accordance with a Manufacturing Technology Transfer Event. Any Confidential Information disclosed by a Party that belongs to a Third Party may be subject to confidentiality obligations that are more stringent that those provided in this Agreement in favor of the Third Party, and in such event the receiving Party shall comply with such additional obligations; provided they are communicated in writing to the receiving Party.

13.2 **Exceptions**. Notwithstanding Section 13.1 (Confidential Information) or anything to the contrary in this Agreement:

13.2.1 **Non-Disclosure Exceptions**. Confidential Information shall not include information and materials (and such information and materials shall not be Confidential Information under this Agreement) that can be established by

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written documentation or other competent proof by the Receiving Party that such information or material: (a) already is in the public domain prior to disclosure by the Disclosing Party or becomes publicly known thereafter through no act, omission or fault of the Receiving Party in breach of this Agreement; (b) is or was already lawfully, and not under an obligation of confidentiality owed to the Disclosing Party under this ARTICLE 13 (Confidentiality), in the possession of the Receiving Party at the time of disclosure by the Disclosing Party; *provided that* the Receiving Party did not initially generate such information and assign its rights to such information to the Disclosing Party in accordance with the terms of this Agreement; (c) is disclosed to the Receiving Party from a Third Party not under an obligation of confidentiality to the Disclosing Party with respect to such information; or (d) has been independently created by the Receiving Party, as evidenced by written or electronic documentation, without any aid, application or use of the Disclosing Party's Confidential Information. Specific aspects or details of Confidential Information will not be deemed to be within the public knowledge or in the prior possession of a Person merely because such aspects or details of the Confidential Information are embraced by general disclosures in the public domain.

13.2.2 **Permitted Disclosures**. The Receiving Party shall have the right to disclose information or materials to the extent required by Applicable Law; *provided that* the Receiving Party (a) uses reasonable efforts to give the Disclosing Party advance notice of such required disclosure in sufficient time to enable the Disclosing Party to seek confidential treatment for such information, (b) provides the Disclosing Party an opportunity to review and comment on such proposed disclosure, and (c) provides reasonable cooperation to assist the Disclosing Party to protect such information and limits the disclosure to that information that is required to be disclosed.

13.3 **Use of Name**. Except as set forth in Section (Use of Corporate Names; Promotional Materials), nothing contained in this Agreement shall be construed as conferring any right to a Party to use in advertising, publicity or other promotional activities any name, trade name, trademark or other designation of the other Party or any of its Affiliates.

13.4 **Publications.**

13.4.1 **In the Regeneron Field**. Subject to the requirements of Section 13.4.3 (Review of Publications), prior to the date of receipt of U.S. Marketing Approval, Regeneron may publish or present the results related to the Product in the Regeneron Field. To the extent such information has not been not previously published or publicly disclosed, Regeneron shall provide Kiniksa with an advance copy of any proposed publication or summary of any proposed oral presentation or poster relating to the Product at least [***] days prior to submission for publication or disclosure for Kiniksa's review and comment.

Kiniksa shall have a reasonable opportunity to recommend any changes it reasonably believes are necessary as set forth in Section 13.4.3 (Review of Publications). Regeneron will give good faith consideration to comments made by Kiniksa with respect to such publications.

13.4.2 **In the Kiniksa Field**. Subject to the requirements of Section 13.4.3 (Review of Publications), Kiniksa may publish or present the results related to the Product in the Kiniksa Field (which for clarity, shall not include the Regeneron Field prior to the date of receipt of U.S. Marketing Approval). To the extent such information has not been not previously published or publicly disclosed, Kiniksa shall provide Regeneron with an advance copy of any proposed publication or summary of any proposed oral presentation or poster relating to the Product at least [***] days prior to submission for publication or disclosure for Regeneron's review and comment. Regeneron shall have a reasonable opportunity to recommend any changes it reasonably believes are necessary as set forth in Section 13.4.3 (Review of Publications). Kiniksa will give good faith consideration to comments made by Regeneron with respect to such publications.

13.4.3 **Review of Publications**. In case of any publication or disclosure provided by the publishing Party to the non-publishing Party for review pursuant to Section 13.4.1 (In the Regeneron Field) or Section 13.4.2 (In the Kiniksa Field), the non-publishing Party shall have the right (a) to propose modifications to the publication or presentation for patent reasons, trade secret reasons, or good faith business reasons, and the publishing Party will remove all Confidential Information of the other Party if requested by the reviewing Party, and (b) to request a reasonable delay in publication or presentation in order to protect patentable information. If the non-publishing Party requests a delay with respect to a patentable invention, then the publishing Party shall delay submission or presentation for a period (not to exceed [***] days) to permit the timely preparation and filing of a Patent Application to protect such Party's rights in such information in accordance with ARTICLE 10 (Intellectual Property).

13.5 **Disclosures Concerning this Agreement**.

13.5.1 **Press Releases**. Subject to Section 13.5.2 (SEC Disclosures), Kiniksa and Regeneron agree not to (and to ensure that their respective Affiliates do not) issue any press releases or public announcements concerning this Agreement or any other activities contemplated hereunder without the prior written consent of the other Party (which consent shall not be unreasonably withheld, conditioned, or delayed). Additionally, the contents of any joint press release or separate press release concerning this Agreement or any other activities contemplated hereunder shall be mutually agreed to between the Parties. Notwithstanding the foregoing, without prior submission to or approval of the other Party, either Party may issue press releases or public announcements that incorporate

information concerning this Agreement or any activities contemplated hereunder to the extent that the information in such release or public announcement has already been publicly disclosed in accordance with the terms of this Agreement and remains correct as of the time of such announcement.

13.5.2 **SEC Disclosures**. Kiniksa acknowledges that Regeneron, as a publicly traded company, is legally obligated to make timely disclosures of all material events relating to its business. Regeneron acknowledges that in the future, Kiniksa may become a publicly traded company, and upon such occurrence, shall be legally obligated to make timely disclosures of all material events relating to its business. Therefore, the Parties acknowledge that either or both Parties may be obligated to issue periodic earnings releases and to make other SEC filings related to the activities contemplated under this Agreement or to file a copy of this Agreement with the U.S. Securities and Exchange Commission or its equivalent in the Territory. Without limiting the generality of the terms of this ARTICLE 13 (Confidentiality), including Section 13.2.2 (Permitted Disclosures), each Party will be entitled to make such filing of this Agreement but shall cooperate with one another and use reasonable efforts to obtain confidential treatment of confidential, including trade secret, information in accordance with Applicable Law. No later than [***] Business Days prior to the anticipated filing, the filing Party will provide the non-filing Party with an advance copy of the Agreement marked to show provisions for which the filing Party intends to seek confidential treatment, allowing a reasonable time for the non-filing Party to review and comment as permitted by Applicable Law, and the filing Party will reasonably consider the non-filing Party's timely comments thereon. In addition, the filing Party will provide the non-filing Party with an advance copy of the securities filings with which the Agreement is furnished or filed or otherwise discussed or disclosed, in each case, only to the extent describing this Agreement, allowing a reasonable time for the non-filing Party to review and comment as permitted by Applicable Law, and the filing Party will reasonably consider the non-filing Party's timely comments thereon; *provided that* the filing Party need not provide for review and comment such securities filings that repeat any such previous disclosures already reviewed and commented upon by the other non-filing Party under the terms of this Section 13.5 (Disclosures Concerning this Agreement) or that contain only non-material factual information regarding this Agreement.

ARTICLE 14
INDEMNITY

14.1 **Indemnity by Kiniksa**. Kiniksa will defend, indemnify and hold harmless Regeneron, its Affiliates and their respective officers, directors, employees, sublicensees and agents ("Regeneron Indemnitees") from and against all losses, liabilities, damages, penalties, fines and expenses, including reasonable attorneys' fees and costs (collectively, "Damages"), arising from or occurring as a result of a Third Party's claim, action, suit,

proceeding (or any judgment, or settlement thereof), in each case, against a Regeneron Indemnitee to the extent it is due to or based upon:

14.1.1 the Development, Commercialization, or the Manufacture of the Product (other than the Manufacture of Non-Conforming Product by Regeneron or its Affiliates) by a Kiniksa Indemnitee in the Kiniksa Field in the Territory in accordance with the license grants in Section 2.1 (Regeneron License to Kiniksa), other than any Development activities in the Kiniksa Field related to any clinical study that Kiniksa is conducting on Regeneron's behalf pursuant to Section 16.8.2(d) (Clinical Studies);

14.1.2 any failure to initiate of recall or market withdrawal of the Product in the Territory, to the extent that Kiniksa is the Recall Responsible Party as of the date of such proposed recall or market withdrawal, and elects not to initiate such a recall or market withdrawal of the Product in the Territory following the Parties' disagreement on whether or not to initiate such a recall or market withdrawal;

14.1.3 the gross negligence or willful misconduct by or of any Kiniksa Indemnitee in performing its or their obligations under this Agreement;

14.1.4 any material breach by Kiniksa of the terms of, or the inaccuracy of any representation or warranty made by it in, this Agreement; or

14.1.5 any claim, litigation, investigation, or proceeding relating to any of the matters specified in Section 12.6 (Compliance with Laws), whether based on contract, tort, or any other theory,

except in each case, Section 14.1.1 to Section 14.1.5, for those Damages for which Regeneron has an obligation to indemnify Kiniksa under Section 14.2 (Indemnity by Regeneron).

14.2 **Indemnity by Regeneron**. Regeneron will defend, indemnify and hold harmless Kiniksa, its Affiliates and their respective officers, directors, employees, sublicensees and agents ("Kiniksa Indemnitees") from and against all Damages arising from or occurring as a result of a Third Party's claim, action, suit, proceeding, (or any judgment, or settlement thereof), in each case, against a Kiniksa Indemnitee to the extent it is due to or based upon:

14.2.1 the Development or Commercialization of the Product by a Regeneron Indemnitee (a) arising prior to the Effective Date, (b) arising after the Effective Date but prior to or on the date of receipt of U.S. Marketing Approval in the Regeneron Field, or (c) otherwise outside the scope of the licenses granted to Kiniksa in Section 2.1 (Regeneron Licenses to Kiniksa), including the Development or Commercialization by a Regeneron Indemnitee in the Regeneron Retained EEO Field;

14.2.2 any Development activities in the Kiniksa Field related to any clinical study that Kiniksa is conducting on Regeneron's behalf pursuant to Section 16.8.2(d) (Clinical Studies);

14.2.3 any Development activities related to Regeneron Exploratory Clinical Studies;

14.2.4 any failure to initiate of recall or market withdrawal of the Product in the Territory, to the extent that Regeneron is the Recall Responsible Party as of the date of such proposed recall or market withdrawal, and elects not to initiate such a recall or market withdrawal of the Product in the Territory following the Parties' disagreement on whether or not to initiate such a recall or market withdrawal;

14.2.5 [***];

14.2.6 the gross negligence or willful misconduct by or of any Regeneron Indemnitee in performing its or their obligations under this Agreement; except that with respect to the gross negligence or willful misconduct by or of any Regeneron Indemnitee in performing its or their obligations under Section 8.9 (Manufacturing Compliance), if such gross negligence or willful misconduct is caused by the Third Party Fill/Finish Provider, then Regeneron's indemnity obligation shall be limited to any amount recovered by Regeneron from the Third Party Fill/Finish Provider in respect of the applicable defect, provided, further, that Regeneron shall use Commercially Reasonable Efforts to engage the Third Party Fill/Finish Provider on commercially reasonable terms, including commercially reasonable indemnities;

14.2.7 any material breach by Regeneron of the terms of, or the inaccuracy of any representation or warranty made by it in, this Agreement; or

14.2.8 any claim, litigation, investigation or proceeding relating to any of the matters specified in Section 12.6 (Compliance with Laws), whether based on contract, tort or any other theory;

except in each case, Section 14.2.1 to Section 14.2.8, for those Damages for which Kiniksa has an obligation to indemnify Kiniksa under Section 14.1 (Indemnity by Kiniksa).

14.3 **Indemnity Procedure.**

14.3.1 **Notice**. The Party entitled to indemnification under this ARTICLE 14 (Indemnity) (an "Indemnified Party") shall notify the Party potentially responsible for such indemnification (the "Indemnifying Party") within [***] Business Days of becoming aware of any claim or claims asserted or threatened in writing against the Indemnified Party that could give rise to a right of indemnification under this Agreement; provided, however, that the failure to give such notice shall not relieve the Indemnifying Party of its indemnity

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obligation hereunder except to the extent that such failure materially prejudices its rights hereunder.

14.3.2 **Defense of Claims**. If the Indemnifying Party has acknowledged in writing to the Indemnified Party the Indemnifying Party's responsibility for defending such claim, the Indemnifying Party shall have the right to defend, at its sole cost and expense, such claim by all appropriate proceedings, which proceedings shall be prosecuted diligently by the Indemnifying Party to a final conclusion or settled at the discretion of the Indemnifying Party; *provided, however*, that the Indemnifying Party may not enter into any compromise or settlement unless (a) such compromise or settlement includes as an unconditional term thereof, the giving by each claimant or plaintiff to the Indemnified Party of a release from all liability in respect of such claim; and (b) the Indemnified Party consents to such compromise or settlement, which consent shall not be withheld, conditioned or delayed unless such compromise or settlement involves (i) any admission of legal wrongdoing by the Indemnified Party, (ii) any payment by the Indemnified Party that is not indemnified hereunder, or (iii) the

imposition of any equitable relief against the Indemnified Party. If the Indemnifying Party does not elect to assume control of the defense of a claim or if a good faith and diligent defense is not being or ceases to be materially conducted by the Indemnifying Party, then the Indemnified Party shall have the right, at the expense of the Indemnifying Party, upon at least ten (10) Business Days' prior written notice to the Indemnifying Party of its intent to do so, to undertake the defense of such claim for the account of the Indemnifying Party (with counsel reasonably selected by the Indemnified Party and approved by the Indemnifying Party, such approval not unreasonably withheld, conditioned, or delayed); *provided that* the Indemnified Party shall keep the Indemnifying Party apprised of all material developments with respect to such claim and promptly provide the Indemnifying Party with copies of all correspondence and documents exchanged by the Indemnified Party and the opposing party to such litigation. The Indemnified Party may not compromise or settle such litigation without the prior written consent of the Indemnifying Party, such consent not to be unreasonably withheld, conditioned, or delayed.

14.3.3 **Participation by the Indemnified Party**. The Indemnified Party may participate in, but not control, any defense or settlement of any claim controlled by the Indemnifying Party pursuant to this Section 14.3 (Indemnity Procedure) and shall bear its own costs and expenses with respect to such participation; *provided*, *however*, [***].

14.4 **Insurance**. During the Term and for a minimum period of [***] years thereafter and for an otherwise longer period as may be required by Applicable Law, each of Regeneron and Kiniksa will (a) procure and maintain appropriate commercial general liability and product liability insurance in an industry-appropriate amounts per occurrence and in the annual aggregate and consistent with [***], or (b) with respect to Regeneron only,

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procure and maintain adequate insurance by means of self-insurance in such amounts and on such terms as are consistent with normal business practices of large pharmaceutical companies in the life sciences industry. Such insurance shall insure against liability arising from this Agreement on the part of Regeneron or Kiniksa, respectively, or any of their respective Affiliates, due to injury, disability, or death of any person or persons, or property damage arising from activities performed in connection with activities performed under this Agreement. Any insurance proceeds received by a Party in connection with any losses shall be retained by such Party and shall not reduce any obligation of the other Party. Each Party shall provide a certificate of insurance or other reasonable documentation evidencing such coverage to the other Party upon its written request. Each Party shall notify the other Party [***] days in advance of cancelation of any such insurance.

ARTICLE 15
FORCE MAJEURE

Neither Party will be held liable or responsible to the other Party nor be deemed to have defaulted under or breached this Agreement for failure or delay in fulfilling or performing any term of this Agreement when such failure or delay is caused by or results from causes beyond the reasonable control of the affected Party (which shall include taking reasonable precautions) including fires, floods, earthquakes, shortages, epidemics, quarantines, embargoes, acts of terrorism, war, acts of war (whether war be declared or not), insurrections, strikes, lockouts, or other labor disturbances, riots, civil commotions, acts of God or acts, omissions, or delays in acting by any Governmental Authority (each, a "Force Majeure"). The affected Party will notify the other Party of such Force Majeure as soon as reasonably practical after such occurrence by giving written notice 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 effective only to the extent of and no longer in duration than the Force Majeure causing the failure or delay in performance and the non-performing Party will use every reasonable effort to remedy its inability to perform due to such Force Majeure.

ARTICLE 16
TERM AND TERMINATION

16.1 **Term**. This Agreement shall begin on the Effective Date and will expire on the date on which neither Kiniksa nor any of its Affiliates, nor any of their respective sublicensees, is Developing or Commercializing any Product in the Kiniksa Field in the Territory under this Agreement (and such cessation of Development and Commercialization activities is acknowledged by Kiniksa in writing to be permanent), unless this Agreement is earlier terminated in its entirety in accordance with this ARTICLE 16 (Term and Termination), in which event the Term shall end on the effective date of such termination.

16.2 **Termination by Kiniksa for Convenience**. Kiniksa may terminate this Agreement at any time commencing on the date that is eighteen (18) months after the Effective Date, (i) upon one hundred eighty (180) days' advanced written notice if such notice is delivered prior to the date of receipt of U.S. Marketing Approval, and (ii) upon one (1) year's

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advanced written notice if such notice is delivered after the date of receipt of U.S. Marketing Approval.

16.3 **Termination for Material Breach**.

16.3.1 **Material Breach**. This Agreement shall be terminable in its entirety by either Party if the other Party commits a material breach of its obligations under this Agreement.

16.3.2 **Notice and Cure**. The terminating Party shall provide the breaching Party with notice of such intended termination, which notice shall set forth in reasonable detail the facts underlying or constituting the alleged material breach (and specifically referencing the provisions of this Agreement alleged to have been breached) and, with respect to alleged material breaches related to non-payment, the termination shall be effective thirty (30) days after the date such notice, and, with respect to all other alleged material breach, the termination shall be effective ninety (90) days after the date such notice is given, unless, in each case, the breaching Party shall have cured such breach within such thirty (30) or ninety (90) day period, as applicable, (or, if such material breach, by its nature, is a curable breach but such breach is not curable within such ninety (90) day period (and for clarity, excluding payment obligations), such longer period as many be required to cure such breach, not to exceed one hundred eighty (180) days, so long as the breaching Party is using reasonable efforts to cure such breach during such period; *provided that* if such breach remains uncured, then such termination shall be effective on the earlier of the expiration of such one hundred eighty (180) day period, or at such time as the breaching Party ceases to use reasonable efforts to cure such breach). Notwithstanding anything to the contrary set forth herein, if either Party reasonably and in good faith disagrees as to whether there has been a material breach, and such Party initiates a dispute resolution procedure under this Agreement to resolve a dispute regarding the alleged material for which termination is being sought and is diligently pursuing such procedure, then the cure period set forth in this Section 16.3 (Termination for Material Breach) will be tolled and will not run during the pendency of such dispute resolution procedures, and in such case all of the terms of this Agreement will remain in effect, and the Parties will continue to perform all of their respective obligations under this Agreement.

16.4 **Termination for Insolvency**. A Party shall have the right to terminate this Agreement in its entirety immediately upon written notice to the other Party if, at any time, (a) the other Party shall file in any court or agency pursuant to any statute or regulation of any state or country a petition in bankruptcy or insolvency or for reorganization or for an arrangement or for the appointment of a receiver or trustee of the other Party or of substantially all of its assets, (b) the other Party shall be served with an involuntary petition filed against it in any insolvency proceeding, and such petition shall not be dismissed within sixty (60) days after the filing thereof, or (c) the other Party is a party to any dissolution or

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liquidation proceedings. In the event that this Agreement is terminated or rejected by a Party or its receiver or trustee under applicable bankruptcy laws due to such Party's bankruptcy, then all rights and licenses granted under or pursuant to this Agreement by such Party to the other Party are, and shall otherwise be deemed to be, for purposes of Section 365(n) of the U.S. Bankruptcy Code and any similar laws in any other country, licenses of rights to "intellectual property" as defined under Section 101(52) of the U.S. Bankruptcy Code. The Parties agree that all Intellectual Property Rights licensed hereunder, including any Patent Rights in any country of a Party covered by the license grants under this Agreement, are part of the "intellectual property" as defined under Section 101(35(A)) of the Bankruptcy Code subject to the protections afforded the non-terminating Party under Section 365(n) of the Bankruptcy Code, and any similar law or regulation in any other country.

16.5 **Termination by Kiniksa for Safety Reasons**. At any time after the Effective Date, Kiniksa may terminate this Agreement on not less than three (3) months' prior written notice to Regeneron (or such earlier period if agreed to by each Party) if Kiniksa reasonably determines based upon its review of the clinical data or upon a determination by an applicable drug safety monitoring board or Governmental Authority (including any action by a Regulatory Authority) that the Product caused or is likely to cause a fatal, life-threatening, or other serious adverse event that is reasonably expected, based upon then-available data, to preclude continued Development or Commercialization of any Product (such termination, a "Safety Termination"). Upon delivery of any such notice of a Safety Termination, Kiniksa may wind-down its then on-going activities related to such Products, including any on-going clinical trials (to the extent consistent with Applicable Law), in accordance with Section 16.8.2(d) (Clinical Studies).

16.6 **Termination for Cessation of Activities**. If at any time during the period commencing on the Effective Date, there is a consecutive twelve (12) month period during which Kiniksa (a) does not conduct any material Development or Commercialization activities for a Product in the Kiniksa Field in the Territory as demonstrated by written and senior management approved budgets and development plans as well as bona fide FTE allocations, or (b) has not granted rights (or an option to obtain rights) to a Third Party to Develop or Commercialize a Product pursuant to a written and executed agreement between Kiniksa and such Third Party (provided that grant of a non-exclusive license to a distributor to sell the Product shall not qualify pursuant to this clause (b)), and such suspension of activity set forth in clauses (a) and (b) is not (i) by written agreement of the Parties, (ii) a result of Kiniksa's reasonable response to guidance from or action or inaction by a Regulatory Authority or other Governmental Authority (such as a clinical hold, a recall, or withdrawal), (iii) a direct result, in whole or in part, of any events outside of the reasonable control of Kiniksa, such as an event of Force Majeure, or any Third Party litigation relating to the Product, then Regeneron may terminate this Agreement with thirty (30) days' written notice to Kiniksa, unless within such thirty (30) day period Kiniksa provides to Regeneron suitable documentation evidencing Kiniksa's conduct of such material Development or Commercialization activities during the applicable twelve (12) month or that Kiniksa is in active and ongoing partnering or

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licensing discussions with a Third Party to grant rights (or an option to obtain rights) to such Third Party to Develop and Commercialize such Product as demonstrated by at least one term sheet exchange between Kiniksa and such Third Party (the last exchange of which has occurred within seventy-five (75) days before the completion such twelve (12) month period). Upon Regeneron's written request to Kiniksa, Kiniksa shall inform Regeneron promptly after receiving such request whether there has been any cessation of material Development or Commercialization activities for a Product in the Kiniksa Field in the Territory and length of such cessation as of the date of such request by Regeneron.

16.7 **Termination for IP Challenge**. If Kiniksa or any of its Affiliates Challenges a Regeneron Patent Right in any country in the Territory (such Patent Right, a "Challenged Patent Right"), then Regeneron may, following written notice to Kiniksa and provided that Kiniksa or its Affiliate does not withdraw such Challenge within thirty (30) days of receipt of such notice, in its sole discretion either (a) exclude such Challenged Patent Right from the scope of the Patent Rights licensed hereunder, or (b) except to the extent the following is unenforceable under the Applicable Law of a particular jurisdiction where a Patent Application within the Challenged Patent Rights is pending or a Patent within the Challenged Patent Rights is issued, terminate this Agreement by providing written notice of termination to Kiniksa. For purposes of this Section 16.7 (Termination for IP Challenge), (i) "Challenge" means, with respect to any Regeneron Patent Right, to contest the validity or enforceability of any such Patent Rights, in whole or in part, in any court, arbitration proceeding or other tribunal, including the United States Patent and Trademark Office and the United States International Trade Commission, and (ii) the term "contest" includes (A) filing an action under 28 U.S.C.§§ 2201-2202 seeking a declaration of invalidity or unenforceability of any such Patent Rights; (B) citation to the United States Patent and Trademark Office pursuant to 35 U.S.C.§ 301 of prior art patents or printed publications or statements of the patent owner concerning the scope of any such Patent Rights; (C) filing a request under 35 U.S.C.§ 302 for re-examination of any such Patent Rights; (D) filing, or joining in, a petition under 35 U.S.C.§ 311 to institute inter partes review of any such Patent Rights or any portion thereof; (E) filing, or joining in, a petition under 35 U.S.C.§ 321 to institute post-grant review of any such Patent Rights or any portion thereof; (F) becoming a party to an interference with an application for any such Patent Rights pursuant to 35 U.S.C.§ 135; (G) any foreign equivalent of clauses (A), (B), (C), (D), (E), or (F) in any country outside the United States; or (G) filing or commencing any opposition, nullity, or similar proceedings challenging the validity of any such Patent Rights in any country; but "Challenge" excludes (1) becoming a party to a Third Party interference for the purpose of defending the validity of any such Patent Rights, (2) filing a request under 35 U.S.C.§ 251 for a reissue of any such Patent Rights, (3) any foreign equivalents of clauses (1) or (2) applicable in any country outside the of United States, (4) the exercise of Kiniksa's rights pursuant to Section 17.18 (Injunctive or Other Equity Relief), (5) any claim as a defense in any lawsuit or administrative proceeding, or (6) any claim or proceeding that would otherwise be a Challenge hereunder to the extent commenced by a Third Party that after the Effective Date acquires or is acquired by Kiniksa or its Affiliates or its or their business or assets, whether by stock purchase, merger, asset purchase, or otherwise;

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provided that such proceeding commenced prior to the closing of such acquisition and further provided that no such Third Party or its advisors has access to Confidential Information of Regeneron and Kiniksa and such Third Party implements reasonable and customary procedures to ensure the foregoing.

16.8 **Effects of Termination or Expiration.**

16.8.1 **All Termination Events**. In the event of any termination of this Agreement (a) except as otherwise provided in this Section 16.8 (Effects of Termination or Expiration), all rights and licenses granted by either Party to the other Party hereunder will immediately terminate and revert to the granting Party, and (b) except as otherwise provided in this Section 16.8 (Effects of Termination or Expiration), all other rights and obligations of the Parties under this Agreement with respect to the Product will terminate.

16.8.2 **Effects of Certain Terminations**. In the event of termination of this Agreement

(a) License Grant to Regeneron. Kiniksa hereby grants to Regeneron an exclusive, (royalty-bearing in accordance with Section 16.8.2(b) (Royalty Payments to Kiniksa) and royalty free and fully-paid up in all other cases), non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing)), worldwide, royalty-free license under the Kiniksa Product Data and a non-exclusive, royalty-free, non-transferable (except as permitted by Section 17.9 (Assignment)), sublicensable through multiple tiers (in accordance with Section 2.7 (Sublicensing), worldwide, royalty-free license under the Kiniksa Platform IP, and Kiniksa's interest in the Joint IP, in each case, to research, develop, make, have made, use, sell, have sold, and import the Product. Any licenses granted by Kiniksa under this Section 16.8.2(a) (License Grant to Regeneron) shall not include the grant of any license under which payments are owed by Kiniksa or its Affiliates to Third Parties on account of the use of such license, unless Regeneron expressly agrees to be responsible for such payments due to such Third Party on account of the use of such license in connection with the research, Development, Commercialization, or Manufacturing of the Product, in which case, such license under the Kiniksa Platform IP will be subject to the scope, terms, and conditions of any Third Party License or other agreement pursuant to which Kiniksa Controls such Kiniksa Platform IP and Regeneron shall assume all obligations to make Third Party License Payments or other payments due under such agreement to the extent that the use of the Kiniksa Platform IP is subject to royalty or other payment obligations in favor of Third Parties.

(b) Royalty Payments to Kiniksa.

(i) *Royalties*. In the event of a termination of this Agreement [***], in consideration for Kiniksa's Development of the Product and the exclusive license granted to Regeneron in Section 16.8.2(a) (License Grant to Regeneron), Regeneron will pay to Kiniksa a royalty of [***] of the aggregate Net Sales of the Product (with the definition of Net Sales applying *mutadis mutandis* to Regeneron or its Affiliates as the selling party) in the Territory. Within [***] days following the end of each Quarter in which such a royalty is owed to Kiniksa for sales of the Product, Regeneron shall provide to Kiniksa a report of the royalties owed (which report will include the information to be provided under Section 9.4.1(a) (Quarterly Net Sales), applied *mutadis mutandis* with Regeneron as the selling party), together with a payment of such royalties owed to Kiniksa for such Quarter. Regeneron shall maintain Records of such sales and royalties for a period of no less than [***] years after Regeneron's obligation to pay such royalties ceases, and Kiniksa shall have the right to audit such Reports, in each case, in accordance with the terms of ARTICLE 11 (Books, Records and Inspections; Audits and Adjustments).

(ii) *Limitation on Royalty Payments*. In the event of a termination of this Agreement by Kiniksa for convenience pursuant to Section 16.2 (Termination by Kiniksa for Convenience), Regeneron's obligation to pay royalties pursuant to Section 16.8.2(b)(i) (Royalties) [***].

(c) Transfer of Promotional Activities. Promptly upon Regeneron's request, Kiniksa shall transfer to Regeneron at Regeneron's direction all Promotional activities related to the Product in the Kiniksa Field in the Territory, including handling of collection and receivables and recording and booking of sales in such country, to Regeneron. In addition, Kiniksa shall provide Regeneron assistance reasonably necessary to ensure an effective transition, including transferring permits and Regulatory Documentation to Regeneron, assisting Regeneron in obtaining new permits and Regulatory Documentation necessary for Regeneron to conduct such Promotional activities, and permitting Regeneron, if permissible under Applicable Law, to use Kiniksa-branded promotional materials during the interim period as the Parties transition the promotional activities.

(d) Clinical Studies. Promptly upon Regeneron's request and at Regeneron's direction, Kiniksa shall either (a) commence the wind down of all clinical studies with respect to the Product in the Kiniksa Field in the Territory, or (b) as soon as reasonably practicable transfer of all clinical study activities with respect to the Product in the Kiniksa Field in the

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Territory to Regeneron; *provided that* in the interim period Kiniksa shall [***], and shall provide Regeneron assistance reasonably necessary to ensure an effective transition, including transferring permits and Regulatory Documentation to Regeneron and assisting Regeneron in obtaining new permits and Regulatory Documentation necessary for Regeneron to conduct such clinical trials.

(e) Transfer of Regulatory Documentation and Approvals; Data. Kiniksa will, as promptly as practicable, and subject to Regeneron's reasonable assistance, to the extent legally permissible (including to the extent permitted under Kiniksa's obligations to Third Parties on the effective date of termination), (a) use reasonable efforts to transfer and assign to Regeneron or Regeneron's designee Kiniksa's rights, title, and interests in and to all material Regulatory Documentation and Approvals (including all BLAs and Pricing Approvals), in all cases, specifically and exclusively relating to the Development, Manufacture, or Commercialization of any Product, and (b) transfer to Regeneron or Regeneron's designee copies of all material Kiniksa Product Data and Product safety data in Regeneron's possession and Control. In the event of (i) any failure to obtain assignment or (ii) with respect to regulatory items that would otherwise fall within (a) or (b), but for such materials not being specifically and exclusively related to a Product, but nonetheless that are necessary for the Development, Manufacture, or Commercialization of any Product, in each of (i) and (ii) Kiniksa hereby consents and grants to Regeneron the right to reference any such item solely with respect to the Development, Commercialization, or the Manufacture of such Product.

(f) Assignment of Third Party Agreements. If Regeneron so requests within [***] after the effective date of termination, Kiniksa will use reasonable efforts, to the extent legally permissible (including to the extent permitted under Kiniksa's obligations to Third Parties on the effective date of termination), to assign to Regeneron any Third Party agreements to which Kiniksa or its Affiliates is a party that are specific to and exclusively relate to the Development, Manufacture, or Commercialization of any Product, subject to any required consents of such Third Party.

(g) Assignment of Product Trademarks. Kiniksa will use reasonable efforts, and subject to Regeneron's reasonable assistance, to the extent legally permissible (including to the extent permitted under Kiniksa's obligations to Third Parties on the effective date of termination), to promptly transfer and assign (or, if applicable, will cause its Affiliates to assign) to Regeneron all of Kiniksa's (and such Affiliates') worldwide rights, title, and interests in and to any Product Trademarks or registered

internet domain names owned by Kiniksa or its Affiliates as of the effective date of termination that are specific to and exclusively used for the Product (it being understood that the foregoing will not include any trademarks or internet domain names that contain the corporate or business names of Kiniksa or any of its Affiliates or any other products of Kiniksa or any of its Affiliates).

(h) Manufacturing Technology Transfer. At Regeneron's request, to the extent that as of the effective date of termination Regeneron has performed a Formulated Bulk Technology Transfer and is no longer Manufacturing such Product, Kiniksa shall perform a Manufacturing technology transfer to Regeneron or Regeneron's designee to the extent necessary to Manufacture such Product in the form Manufactured by or on behalf of Kiniksa as of the effective date of termination.

(i) Supply to Regeneron. If, as of the effective date of termination, Kiniksa is Manufacturing a Product and Regeneron is no longer Manufacturing such Product, then, at Regeneron's request and only for the period of time reasonably necessary to ensure patients have access to the Product, such period not to exceed [***] after the initiation of a Manufacturing technology transfer pursuant to Section 16.8.2(h) (Manufacturing Technology Transfer), Kiniksa shall use reasonable efforts to continue to Manufacture such Product on behalf of Regeneron and its Affiliates and licensees, in each case, at a purchase price equal to Kiniksa's Fully-Burdened Cost.

(j) Transfer of Prosecution and Maintenance Responsibilities. Kiniksa shall transfer to Regeneron any and all Regeneron Patent Right prosecution and maintenance responsibilities, including transferring all files related to the prosecution and maintenance of such Regeneron Patent Rights, and at the request of Regeneron, Kiniksa shall make appropriate personnel available to Regeneron to answer such reasonable questions as Regeneron may have in connection with such transfer of prosecution and maintenance of such Patent Rights.

(k) Kiniksa Sell-Off Right. Notwithstanding anything to the contrary set forth herein, except if this Agreement is terminated by Regeneron pursuant to Section 16.3 (Termination for Material Breach), Section 16.4 (Termination for Insolvency), Section 16.6 (Termination for Cessation of Activities) or Section 16.7 (Termination for IP Challenge) or this Agreement is terminated by Kiniksa pursuant to Section 16.2 (Termination by Kiniksa for Convenience) or Section 16.5 (Termination by Kiniksa for Safety Reasons), the licenses granted to Kiniksa under this Agreement shall survive for a period of [***] after the effective date of termination of this Agreement, solely to the limited extent necessary

to enable Kiniksa (and its Affiliates and sublicensees) to, at their discretion, sell-off any Products in accordance with this Agreement then remaining in its or its Affiliates' existing inventory or that are works-in-process as of the effective date of termination. The Profit Split Arrangement, including all sharing of Other Shared Expenses, Shared Commercial Expenses, and COGS, in each case, shall apply with respect to any Product sold by Kiniksa pursuant to the Product sell-off under this Section 16.8.2(k) (Kiniksa Sell-Off Right). If this Agreement is terminated by Regeneron pursuant to Section 16.3 (Termination for Material Breach), Section 16.4 (Termination for Insolvency), Section 16.6 (Termination for Cessation of Activities) or Section 16.7 (Termination for IP Challenge) or this Agreement is terminated by Kiniksa pursuant to Section 16.2 (Termination by Kiniksa for Convenience) or Section 16.5 (Termination by Kiniksa for Safety Reasons), at Regeneron's request and direction, Kiniksa shall ship all Product inventory to Regeneron.

(l) Responsibility for Costs. Within [***] after receipt of an invoice therefor, Regeneron will reimburse Kiniksa the reasonable costs incurred by Kiniksa in connection with Kiniksa's performance of activities under this Section 16.8 (Effects of Termination or Expiration); *provided that* (a) [***], and (b) [***] incurred by Kiniksa in connection with its performance of activities under this Section 16.8 (Effects of Termination or Expiration).

16.9 **Confidentiality**. In the event of a termination of this Agreement, if there is Confidential Information of both Parties relating specifically to the Product, then such Confidential Information shall become Confidential Information of Regeneron, but for clarity Kiniksa shall have the rights set forth in Section 13.2.2 (Permitted Disclosures).

16.10 **Survival of Obligations**. Except as otherwise provided below, upon termination of this Agreement, the rights and obligations of each Party hereunder shall terminate, and this Agreement shall cease to be of further force or effect:

16.10.1 **Accrued Rights**. neither Kiniksa nor Regeneron shall be relieved of any obligations (including payment obligations) of such Party arising prior to termination, including the payment of any non-cancelable costs and expenses i (even if such costs and expenses arise following termination or expiration, as the case may be); and

16.10.2 **Surviving Sections**. The obligations of the Parties set forth in Articles ARTICLE 1, ARTICLE 11, ARTICLE 13, ARTICLE 14, ARTICLE 17 and Section 2.7.3 (Continuation of Sublicenses Upon Termination), Section 9.3 (Sharing of Third Party Proceeds), Section 9.4 (Sharing of Profits, Periodic Reports, and Fund Flow Mechanics), Section 9.5 [***] Inclusion of Certain Shared Commercial Expenses), Section 9.5.2 (Certain Shared Commercial

Expenses Cost True-Up), Section 9.7 (Late Payments), Section 9.8 (Taxes), Section 16.8 (Effects of Termination or Expiration), Section 16.9 (Confidentiality), and Section 16.10 (Survival of Obligations), shall survive and expiration or termination of this Agreement and continue to be enforceable.

16.11 **Return of Confidential Information**. Confidential Information disclosed by the Disclosing Party, including permitted copies, shall remain the property of the Disclosing Party. Subject to the principles set forth in Section 16.9 (Confidentiality), upon the termination of this Agreement, or the written request of the Disclosing Party, the Receiving Party shall promptly return to the Disclosing Party or, at the Disclosing Party's request, destroy, all documents or other tangible materials embodying the Disclosing Party's Confidential Information (or any designated portion thereof); provided that (a) copies may be maintained in the confidential files of the Receiving Party for the purpose of complying with the terms of this Agreement, and (b) the Receiving Party may retain the Disclosing Party's Confidential Information that is reasonably necessary for the practice of any license from the Disclosing Party to the Receiving Party that survives termination of this Agreement, as applicable. The Receiving Party also shall certify in writing that it has satisfied its obligations under this Section 16.11 (Return of Confidential Information) within [***] days of a written request by the Disclosing Party.

ARTICLE 17
MISCELLANEOUS

17.1 **Disputes.**

17.1.1 **Governing Law; Submission to Jurisdiction**. This Agreement shall be governed by and construed in accordance with the laws of the State of New York, without regard to the conflict of laws principles thereof that would require the application of the law of any other jurisdiction. Except for Financial Disputes, which are governed by Section 17.1.2 (Financial Disputes), the Parties irrevocably and unconditionally submit to the exclusive jurisdiction of the United States District Court for the Southern District of New York solely and specifically for the purposes of any action or proceeding arising out of or in connection with this Agreement.

17.1.2 **Financial Disputes**. Financial Disputes shall be submitted for resolution to a Big Four accounting firm jointly selected by each Party's certified public accountants or to such other Person as the Parties shall agree (the "Financial Expert"). Each Party shall submit its position or proposal on the applicable matter and a brief supporting such proposal or position within twenty (20) Business Days after selection of the Financial Expert. The Financial Expert will make its determination within [***] days after its receipt of each Party's brief to be providing pursuant to the foregoing sentence; *provided that* with respect to [***]. The decision of the Financial Expert shall be final and the costs of the Financial Expert shall be borne by the Parties in accordance with such allocation as the Financial Expert shall determine.

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.

17.2 **Waiver**. The failure of a Party to insist on the performance of any obligation hereunder by the other Party shall not be deemed to be a waiver of such obligation. Waiver by a Party of a breach hereunder by the other Party shall not be construed as a waiver of any subsequent breach of the same or any other provision. No delay or omission by a Party in exercising or availing itself of any right, power, or privilege hereunder shall impair, prejudice, or constitute a waiver of or preclude the later exercise of any such right, power or privilege by such Party. No waiver shall be valid or effective unless made in writing with specific reference to the relevant provision of this Agreement and signed by a duly authorized representative of each Party.

17.3 **Notices**. All notices, instructions, and other communications required or permitted hereunder or in connection herewith shall be in writing, shall be sent to the address of the relevant Party set forth on Schedule 17.3 attached hereto and shall be (a) delivered personally, (b) sent via a reputable nationwide overnight courier service, or (c) sent by facsimile transmission, with a confirmation copy to be sent by registered or certified mail, return receipt requested, postage prepaid, except in the event this Agreement specifies the notice may be delivered by email. Any such notice, instruction, or communication shall be deemed to have been delivered: (a) upon receipt if delivered by hand, (b) one (1) Business Day after it is sent via a reputable nationwide overnight courier service, or (c) when transmitted with electronic confirmation of receipt, if transmitted by facsimile (or email, if email is permitted), if such transmission is made during regular business hours of the recipient on a Business Day; or otherwise, on the next Business Day following such transmission. Either Party may change its address by giving written notice to the other Party in the manner provided above.

17.4 **Entire Agreement**. This Agreement contains the complete understanding of the Parties with respect to the subject matter hereof, and supersedes all prior understandings and writings relating to the subject matter hereof, whether written or oral. For clarity, this Agreement supersedes the CDA; provided that all confidential or proprietary information exchanged by the Parties pursuant to the CDA will be Confidential Information for purposes of this Agreement. In the event of any conflict between this Agreement and the Pharmacovigilance Agreement or any Supply Agreement or Quality Agreement, the terms of this Agreement shall control unless expressly set forth to the contrary in any such Pharmacovigilance Agreement (solely with respect to safety-related matters) in which case the Pharmacovigilance Agreement shall control, Supply Agreement (solely with respect to the Manufacture and supply of Product) in which case the Supply Agreement shall control, or Quality Agreement (with respect to Product quality-related matters) in which case the Quality Agreement shall control.

17.5 **Amendments**. No provision in this Agreement (or in any schedule or exhibit) shall be supplemented, deleted, or amended except in a writing executed by an authorized representative of each of Kiniksa and Regeneron.

17.6 **Interpretation**. The captions to the several Articles and Sections of this Agreement are included only for convenience of reference and shall not in any way affect the

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construction of, or be taken into consideration in interpreting, this Agreement. In this Agreement: (a) the word "including" shall be deemed to be followed by the phrase "without limitation" or like expression; (b) references to the singular shall include the plural and vice versa; (c) references to masculine, feminine and neuter pronouns and expressions shall be interchangeable; (d) the words "herein" or "hereunder" relate to this Agreement; (e) the words "shall" and "will" have the same meaning; (f) references to a particular statute or regulation include all rules and regulations thereunder and any predecessor or successor statute, rules or regulation, in each case as amended or otherwise modified from time to time; (g) words in the singular or plural form include the plural and singular form, respectively; (h) references to a particular person include such person's successors and assigns to the extent not prohibited by this Agreement; (i) unless otherwise specified, "$" is in reference to United States dollars; and (j) the word "or" has the inclusive meaning represented by the phrase "or/and." Each accounting term used herein that is not specifically defined herein shall have the meaning given to it under GAAP, but only to the extent consistent with its usage and the other definitions in this Agreement.

17.7 **Construction**. The Parties acknowledge and agree that: (a) each Party and its counsel reviewed and negotiated the terms and provisions of this Agreement and have contributed to its revision; (b) the rule of construction to the effect that any ambiguities are resolved against the drafting Party will not be employed in the interpretation of this Agreement; and (c) the terms and provisions of this Agreement will be construed fairly as to each Party and not in a favor of or against either Party, regardless of which Party was generally responsible for the preparation of this Agreement. The headings of clauses contained in this Agreement preceding the text of the sections, subsections, and paragraphs hereof are inserted solely for convenience and ease of reference only and shall not constitute any part of this Agreement, or have any effect on its interpretation or construction. This Agreement has been prepared in the English language and the English language shall control its interpretation. In addition, all notices required or permitted to be given hereunder, and all written, electronic, oral, or other communications between the Parties regarding this Agreement shall be in the English language.

17.8 **Severability**. If, under Applicable Laws, any provision hereof should be held or otherwise determined to be invalid, illegal, or unenforceable in any jurisdiction, then the Parties shall agree upon valid and enforceable provisions ("Modified Clause") for such invalid, illegal, or unenforceable provisions, which valid provisions in their economic effect are sufficiently similar to the invalid, illegal, or unenforceable provisions that it can be reasonably assumed that the Parties would have entered into this Agreement with such valid provisions. This Agreement shall endure and that the Modified Clause shall be enforced in such jurisdiction to the maximum extent permitted under Applicable Laws in such jurisdiction; provided that the Parties shall consult and use all reasonable efforts to agree upon, and hereby consent to, any valid and enforceable modification of this Agreement as may be necessary to avoid any unjust enrichment of either Party and to match the intent of this Agreement as closely as possible, including the economic benefits and rights contemplated herein.

17.9 **Assignment.**

17.9.1 **Assignment to Third Parties by Kiniksa**. Prior to the date of receipt of U.S. Marketing Approval, Kiniksa may not assign its rights or obligations under this Agreement to any Third Parties without Regeneron's consent. After the date of receipt of U.S. Marketing Approval, Kiniksa may assign its rights and obligations under this Agreement to a Third Party upon advance written notification to Regeneron setting forth the name of assignee and subject to Section 2.11 (Regeneron ROFN to Purchase Rights to Product). Regeneron's consent with respect to the assignee shall not be required, unless (a) [***], (b) [***] pursuant to this Section 17.9.1 (Assignment to Third Parties by Kiniksa), [***]. If any of clauses (a), (b), or (c) in the previous sentence applies, Regeneron's consent shall be required and within [***] days after receiving notice pursuant to this Section 17.9.1 (Assignment to Third Parties by Kiniksa), Regeneron shall inform Kiniksa in writing as to whether any of clauses (a), (b), or (c) applies. With respect to any communications between the Parties with respect to clause (c), the Parties shall cooperate in good faith as to agree to procedures that would maintain the privileged nature of such information. For clarity, this Section 17.9.1 (Assignment to Third Parties by Kiniksa) shall not apply to a Change of Control of Kiniksa, which shall be covered by Section 2.12 (Change of Control of Kiniksa).

17.9.2 **Assignment to Affiliates by Kiniksa**. At all times, Kiniksa may assign its rights and obligations under this Agreement to its Affiliate without Regeneron's written consent that has and will continue to have the resources and financial wherewithal to fully meet its obligations under this Agreement.

17.9.3 **Assignment by Regeneron**. Regeneron shall not require Kiniksa's consent to assign its rights and obligations under this Agreement.

17.9.4 **Assignee Obligations**. Any Affiliate or Third Party assignee, as a condition of such assignment shall agree in writing to be bound by the terms of this Agreement.

17.9.5 **Invalid Assignments**. Any attempted assignment in violation of this Section 17.9 (Assignment) and Section 2.11 (Regeneron ROFN to Purchase Rights to Product) shall be void.

17.10 **Guarantee**. Kiniksa Pharmaceuticals Corp., a Delaware corporation and a subsidiary of Kiniksa ("Kiniksa Corp"), hereby unconditionally guarantees the obligations of Kiniksa under this Agreement. In the event of any claim or cause of action by Regeneron under this Agreement, Kiniksa and Kiniksa Corp each acknowledge and agree that Regeneron may bring such claim or cause of action directly against Kiniksa Corp without an obligation to first bring such claim or cause of action against Kiniksa or to first exhaust any of its rights against Kiniksa.

17.11 **Successors and Assigns**. This Agreement shall be binding upon and inure to the benefit of the Parties and their respective successors and permitted assigns. Except as expressly set forth in this Agreement, no person or entity other than the Parties and their respective Affiliates and permitted assignees hereunder shall be deemed an intended beneficiary hereunder or have any right to enforce any obligation of this Agreement.

17.12 **Performance by Affiliates**. Each Party acknowledges and accepts that the other Party may exercise its rights and perform its obligations under this Agreement either directly or through one or more of its Affiliates. Without limiting the foregoing, neither Party shall cause or permit any of its Affiliates to commit any act (including any act or omission) which such Party is prohibited hereunder from committing directly. A Party's Affiliates will have the benefit of all rights (including all licenses) of such Party under this Agreement where necessary to give such Party's Affiliates the benefits of the rights provided to such Party in this Agreement, provided that each Party will remain responsible for the acts and omissions, including any financial liabilities, of its Affiliates.

17.13 **Further Assurances and Transaction Approvals**. Upon the terms and subject to the conditions hereof, each of the Parties will (a) take, or cause to be taken, all actions necessary, proper or advisable under Applicable Laws or otherwise to consummate and make effective the transactions contemplated by this Agreement, (b) obtain from the requisite Governmental Authorities any consents, licenses, permits, waivers, approvals, authorizations, or orders required to be obtained or made in connection with the authorization, execution, and delivery of this Agreement and the consummation of the transactions contemplated by this Agreement, and (c) make all necessary filings, and thereafter make any other advisable submissions, with respect to this Agreement and the transactions contemplated by this Agreement required under Applicable Laws.

17.14 **Counterparts**. This Agreement may be executed in counterparts, each of which shall be deemed an original, shall be construed together and shall constitute one and the same instrument.

17.15 **No Third Party Beneficiaries**. Except as expressly provided in ARTICLE 14 (Indemnity) with respect to the Regeneron Indemnitees or Kiniksa Indemnitees, none of the provisions of this Agreement shall be for the benefit of or enforceable by any Third Party, including any creditor of any Party hereto. No Third Party shall obtain any right under any provision of this Agreement or shall by reason of any such provision make any claim in respect of any debt, liability or obligation (or otherwise) against any Party hereto.

17.16 **Relationship of the Parties**. Each Party shall bear its own costs incurred in the performance of its obligations hereunder without charge or expense to the other Party except as expressly provided in this Agreement. Neither Kiniksa nor Regeneron shall have any responsibility for the hiring, termination or compensation of the other Party's employees or for any employee compensation or benefits of the other Party's employees. No employee or representative of a Party shall have any authority to bind or obligate the other Party to this Agreement for any sum or in any manner whatsoever, or to create or

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impose any contractual or other liability on the other Party without said Party's approval. For all purposes, and notwithstanding any other provision of this Agreement to the contrary, Regeneron's legal relationship under this Agreement to Kiniksa, and Kiniksa's legal relationship under this Agreement to Regeneron, shall be that of an independent contractor. Nothing in this Agreement shall be construed to establish an employment, agency, joint venture, or partnership between the Parties or any of their respective Affiliates. For purposes of this Agreement, as of the Effective Date, neither Kiniksa nor any of its Affiliates is an Affiliate of Regeneron or any of its Affiliates, and neither Regeneron nor any of its Affiliates is an Affiliate of Kiniksa or any of its Affiliates.

17.17 **Limitation of Liability**. NEITHER REGENERON NOR KINIKSA SHALL BE LIABLE FOR SPECIAL, PUNITIVE, INDIRECT, INCIDENTAL OR CONSEQUENTIAL DAMAGES ARISING OUT OF THIS AGREEMENT OR THE EXERCISE OF ITS RIGHTS HEREUNDER, INCLUDING LOSS OF PROFITS ARISING FROM OR RELATING TO ANY BREACH OF THIS AGREEMENT, REGARDLESS OF THE THEORY OF LIABILITY (INCLUDING CONTRACT, TORT, NEGLIGENCE, STRICT LIABILITY OR OTHERWISE) AND REGARDLESS OF ANY PRIOR NOTICE OF SUCH DAMAGES, EXCEPT AS A RESULT OF A PARTY'S WILLFUL MISCONDUCT, A BREACH OF THE CONFIDENTIALITY AND NON-USE OBLIGATIONS IN ARTICLE SECTION 13.1 (CONFIDENTIAL INFORMATION).

NOTHING IN THIS SECTION 17.17 (LIMITATION OF DAMAGES) IS INTENDED TO LIMIT OR RESTRICT THE INDEMNIFICATION RIGHTS OR OBLIGATIONS OF EITHER PARTY HEREUNDER WITH RESPECT TO THIRD PARTY CLAIMS.

17.18 **Injunctive or Other Equity Relief**. 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.

17.19 **Rights in Bankruptcy**. The Parties agree that all licenses and rights to licenses granted under or pursuant to this Agreement by Regeneron or Kiniksa are and shall otherwise be deemed to be, for purposes of Section 365(n) of the Bankruptcy Code or any analogous provisions in any other country or jurisdiction, including any Patent Rights in any country of a Party covered by the license grants under this Agreement, licenses of rights to "intellectual property" as defined under Section 101(35(A)) of the U.S. Bankruptcy Code subject to the protections afforded the non-bankrupt Party under Section 365(n) of the Bankruptcy Code, and any similar law or regulation in any other country. 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

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non-subject Party's possession, shall be promptly delivered to it (and such access shall be provided promptly) (a) 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 (b) if not delivered under clause (a) 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.

17.20 **Non-Exclusive Remedies**. 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 and to the extent expressly set forth herein (including as expressly set forth in Section 8.11 (Limitation of Damages)).

[Remainder of page intentionally left blank; Signature page follows]

IN WITNESS WHEREOF, Regeneron and Kiniksa have caused this Agreement to be executed by their duly authorized representatives as of the Effective Date.

REGENERON PHARMACEUTICALS, INC.

By /s/ Leonard Schleifer
Name: Leonard Schleifer
Title: President and CEO

KINIKSA PHARMACEUTICALS, LTD.

By /s/ Thomas Beetham
Name: Thomas Beetham
Title: Executive Vice President

Acknowledged and Agreed (solely with respect to Section 17.10 (Guarantee)):

KINIKSA PHARMACEUTICALS CORP.

By /s/ Chris Heberlig
Name: Chris Heberlig
Title: Chief Financial Officer

[Signature Page to License Agreement]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 1.58

Existing Product Trademarks

[***]

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.

SCHEDULE 1.165

Regeneron Patent Rights

[See attached]

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.

[***]

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.

SCHEDULE 1.179

Retained Field

Indications:

(i) Cryopyrin-associated periodic syndrome (CAPS), (ii) [***] and (iii) "Other Indications" medically related to [***].

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.

SCHEDULE 1.181

Retained Territory

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 3.5.2

Existing Contracts in the Regeneron Field, all IIS Studies in the Territory and Regeneron Exploratory Clinical Studies in the Kiniksa Field in the Territory

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 5.4

Initial Development Plan

[See Attached]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 8.3

Development Supply Agreement Term Sheet

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 9.4

Profit Share Arrangement

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 10.7.4

Planned Patent Applications

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 10.13.1

Trademark Co-Existence Agreements

[***]

Confidential Treatment Requested by Kiniksa Pharmaceuticals, Ltd.

SCHEDULE 17.3

Notices

If to Regeneron:

Regeneron Pharmaceuticals, Inc.
777 Old Saw Mill River Road
Tarrytown, New York 10591
Attention: President & CEO
Copy: General Counsel

If to Kiniksa:

Kiniksa Pharmaceuticals, Ltd.
Clarendon House
2 Church Street
Hamilton HM 11
Bermuda
Attention: Chief Legal Officer

With a copy to:
Kiniksa Pharmaceuticals Corp.
15 Walnut Street
Wellesley, Massachusetts 02481
Attention: Legal Department

With a copy to (which shall not constitute notice for purposes of this Agreement):
Ropes & Gray LLP
Prudential Tower, 800 Boylston Street
Boston, Massachusetts 02199
Attention: David M. McIntosh

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.

Exhibit 10.12

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.

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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.

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

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(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

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

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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 [***] 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

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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

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

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

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.

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

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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 Subliensee'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

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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

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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

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.

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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

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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

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.

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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(v) 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,

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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

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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

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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

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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

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.

Schedule 1.39

[***]

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.

Schedule 1.56

[***]

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.

Schedule 1.74

[***]

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.

Schedule 1.104
Transition Activities

[***]

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.

Schedule 3.1 Inventory

[***]

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.

Schedule 8.2.3

[***]

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.

CLINICAL SUPPLY AGREEMENT

THIS CLINICAL SUPPLY AGREEMENT ("**Agreement**") effective as of September 27, 2017 ("**Effective Date**"), is by and between Regeneron Pharmaceuticals, Inc. ("**Regeneron**"), and Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company ("**KINIKSA"**). Regeneron and KINIKSA are sometimes hereinafter referred to as a "**Party**" and collectively as the "**Parties**".

WHEREAS, Regeneron and KINIKSA are Parties to that certain License Agreement, dated as of September 25, 2017 ("**LA**"), pursuant to which KINIKSA will be engaged in the Development of the Product in the Kiniksa Field in the Territory and Regeneron will Manufacture and supply Product to KINIKSA for such purposes; and

WHEREAS, pursuant to the terms of the LA, Regeneron intends to Manufacture and supply Formulated Bulk Product, Filled Product, and packaged and labeled Product (if applicable) and placebo for Clinical Supply Requirements pursuant to the LA;

NOW, THEREFORE, in consideration of the foregoing, of the mutual covenants and undertakings contained herein and of other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, the Parties, intending to be legally bound, hereby agree as follows.

1. Definitions.

Capitalized terms used herein shall have the defined meanings set forth in the LA, unless otherwise defined herein.

2. Agreement to Supply; Forecasts; Purchase Orders; Deviating Product.

2.1 Generally. Subject to the terms and conditions of this Agreement, during the Term, Regeneron shall manufacture, fill, package and, if applicable, label (the content of any such labeling and packaging to be made in accordance with KINIKSA's reasonable instructions) and supply to KINIKSA the Formulated Bulk Product or Filled Product (either, as applicable, "**Finished Product**") and placebo solely for use in Development in the Kiniksa Field in the Territory, all in accordance with the terms of the LA and this Agreement, and in such form of the Product as set forth in Forecasts and Firm Orders (i.e., whether the Product will be in the form of Formulated Bulk Product, Filled Product or packaged and labeled Product, or placebo). In accordance with, and to the extent set forth in, the LA and the process set forth in the Quality Agreement, Regeneron may transfer a portion or all of such activities to one or more Third Parties or Affiliates. To the extent KINIKSA desires for REGENERON to provide packaging and labeling for Filled Product, KINIKSA shall, within the timeframe agreed upon by the Parties, be responsible for providing Regeneron (or its designee) with informational content for all labels and packaging (including package inserts) for the Product and placebo for use in the Kiniksa Field Territory, and KINIKSA shall be solely responsible to ensure that such content complies with applicable Laws. Title and risk of loss for Finished Product and placebo shall be in accordance with Incoterms 2010, EXW. After the completion of a successful Fill/Finish Technology Transfer in accordance with the LA, Regeneron's sole obligation shall be to supply Formulated Bulk Product under this Agreement.

2.2 Forecasts and Purchase Orders.

(a) Subject to Section 2.2(b), KINIKSA shall provide Regeneron in writing with a forecast of quantities and delivery dates for the requirements of the Finished Product and placebo to be supplied under this Agreement (each a "**Forecast**") no later than thirty six (36) months prior to the first requested delivery date and, thereafter, KINIKSA shall provide Regeneron with updated, rolling, thirty-six (36) month Forecasts on a quarterly basis.

(b) The Parties acknowledge that it may be difficult for KINIKSA to provide a Forecast meeting the required lead time set forth in this Section for its first pilot clinical trial and first pivotal clinical study in the Territory (together, the "**Initial Clinical Studies**"); and, accordingly and notwithstanding the foregoing, KINIKSA shall provide a Forecast for such study with as much lead time as reasonably possible. Regeneron shall consult and coordinate with KINISKSA regarding the feasibility of fulfilling said Forecast and shall make commercially reasonable efforts to fulfill said Forecast.

(c) Promptly following receipt of a Forecast, Regeneron shall notify KINIKSA of its ability to supply the requirements of the Forecast. In the event Regeneron notifies KINIKSA that it is able to meet such requirements, then such Forecast shall be deemed accepted by Regeneron. On the other hand, if Regeneron notifies KINIKSA that it is not able to satisfy a Forecast, then Regeneron shall prepare and provide KINIKSA with a time schedule for additional Manufacturing of the Finished Product and/or placebo to satisfy the requirements of such Forecast within (10) Business Days of receipt of such Forecast, and (i) the Parties shall mutually agree upon a revised Forecast consistent with such time schedule, and (ii) upon such mutual agreement, such revised Forecast shall be deemed accepted by Regeneron. Regeneron shall not be obligated to supply Product or placebo under this Agreement except pursuant to a Forecast that is accepted by Regeneron in accordance with this Section 2.2.

(d) An accepted Forecast may be modified by KINIKSA to increase or decrease the number of units of Finished Product and/or placebo to be supplied on a certain delivery date under such Forecast, solely as follows:

(i) during the time period occurring more than twelve (12) months prior to such delivery date, KINIKSA may modify the number of such units without restriction, and;

(ii) during the time period occurring within twelve (12) months prior to such delivery date ("**Firm Order Commitment**"), KINIKSA may not modify the number of such units, without Regeneron's prior written consent.

(e) KINIKSA shall provide Regeneron with firm purchase orders for Finished Product and/or placebo at least twelve (12) months prior to the delivery dates specified in an accepted Forecast ("**Firm Orders**"). Each Firm Order submitted will be consistent with the Firm Order Commitment. If Kiniksa fails to meet its Firm Order Commitment, then Regeneron will use reasonable efforts to reallocate the excess Product. If Regeneron cannot re-allocate any such Product, then Kiniksa will be obligated to purchase such Product. If Kiniksa's orders exceed the firm order forecast, then Regeneron will use reasonable efforts to meet those orders, but will not be liable if it is not able to do so.

(f) Finished Product shall have a shelf life of at least twenty-four (24) months ("**Minimum Shelf Life**") at the time of receipt by Kiniksa (or its designee); provided that Kiniksa acknowledges that Finished Product for the Initial Clinical Studies may not have the Minimum Shelf Life, in which case it shall have the shelf life agreed upon by the Parties at time that Kiniksa submits a purchase order for such Finished Product.

(g) All Firm Orders for Finished Product (to the extent not already finished and on hand) must be in multiples of 1250 vials.

2.3 Production Requirements. Subject to the terms of this Agreement and the LA, Regeneron shall apply its production capacity that, in Regeneron's reasonable discretion, is necessary to fulfill the quantity and delivery date requirements set forth in each Firm Order; provided such Firm Order is consistent with an accepted Forecast.

2.4 Deviating Product.

(a) If Formulated Bulk Product or Filled Product or placebo is released by Regeneron, is delivered to KINIKSA and inspected by Kiniksa (or its agents) and deviates from Specifications (as defined below) and such deviation was attributable to Regeneron ("**Deviating Product/Placebo**"), Regeneron will, as soon as commercially practical, replace the Deviating Product/Placebo with conforming Finished Product or placebo, as the case may be. Regeneron will bear the cost of any replacement Finished Product and/or placebo and the transportation, testing and disposal costs of any Deviating Product/Placebo. For clarity, Deviating Product/Placebo does not include Failed Batches.

(b) "**Failed Batch**" means any batch of Product Manufactured by Regeneron or the Third Party Fill/Finish Provider that fails to generate adequate yields of Product or fails to meet quality requirements, including failure to Manufacture any Product in compliance with Specifications or applicable Good Practices, in each case which were not released by Regeneron. If a Failed Batch does not result from the gross negligence, recklessness, bad faith or intentional wrongful acts, or omissions committed by Regeneron or its Affiliates (or their respective agents, contractors, representatives or other persons or entities working on their behalf), then, to the extent such Failed Batch was Manufactured, in whole or in part to fulfill Kiniksa's Firm Order, the costs of the Failed Batch (or a pro rata allocation thereof if the Failed Batch was not Manufactured solely to fulfill Kiniksa's Firm Order) will be included as Manufacturing Costs of Product as set forth in the LA (in the definition of Manufacturing Costs), otherwise KINIKSA shall not be responsible for the costs of such Failed Batch.

3. Supply Price of the Product for use in Development.

3.1 Supply Price. The supply price of the Finished Product (in whatever form applicable) and placebo to be supplied by Regeneron under this Agreement shall be at the rate determined as set forth in the LA.

3.2 Payment Method and Currency. All payments under this Agreement shall be made pursuant to and be applied as set forth in the LA.

4. Quality Control; Conflict Between Agreements.

4.1 Quality Agreement. The Parties (or their Affiliates) shall enter into a separate quality agreement with respect to the quality assurance, filling, packaging and labeling of Finished Product and placebo supplied by Regeneron hereunder ("**Quality Agreement**"). In the event there is any conflict relating to quality control procedures or cGMP-related activities between the terms and provisions of this Agreement and the Quality Agreement, the applicable terms and provisions of the Quality Agreement shall control and prevail with respect to matters specifically related to quality assurance, filling, packaging and labeling of Finished Product and placebo; provided that, with respect to all other matters, the terms, provisions and conditions of this Agreement or LA shall control and prevail. In the event of a conflict in provisions between this Agreement and LA, the LA shall control and prevail, except solely with respect to the supply and Manufacture of Product, in which case this Agreement shall control.

5. Warranties; Limitation of Liability.

5.1 Warranty and Limitation of Liability. In addition to specific warranties and any limitation of liability included herein, warranties and limitation of liability of Regeneron and KINIKSA shall be governed under the LA.

5.2 General Warranties. Regeneron represents and warrants that (a) the applicable Finished Product and placebo supplied under this Agreement shall be Manufactured, stored, processed, packaged, labeled and tested in accordance with the specifications provided by Regeneron under and in accordance with the Quality Agreement ("**Specifications**") and all Applicable Laws; (b) applicable Finished Product and placebo furnished by Regeneron to KINIKSA under this Agreement (i) shall be free of any defects in any materials or workmanship, (ii) shall be stored and supplied in conformity with the Specifications and all Applicable Laws, and (iii) shall not contain any material provided by or on behalf of Regeneron that has not been used or stored in accordance with the Specifications and Applicable Laws; (c) it will not introduce any materials not provided in the Specifications that would cause the applicable Finished Product or placebo to be adulterated within the meaning of Section 501 of the U.S. Federal Food, Drug and Cosmetics Act and the regulations promulgated thereunder; and (d) it shall perform all obligations hereunder in compliance with all Applicable Laws and industry standards of workmanship and professionalism.

5.3 Debarment. Section 12.4.11 of the LA is hereby incorporated into a made a part of this Agreement.

6. Term; Termination.

The term of this Agreement shall be from the Effective Date until the earlier of the completion of a Formulated Bulk Manufacturing Technology Transfer or termination of the LA.

7. Confidentiality.

Confidentiality shall be governed under the LA (references in the LA to the LA (e.g. "this Agreement", hereunder, or similar references) in the confidentiality provisions and related definitions shall be deemed to also refer to this Agreement for purposes of this Section 7).

8. Miscellaneous.

8.1 Authority to Enter Into Agreement. Each Party represents and warrants that it is authorized to enter into this Agreement and that in so doing it is not in violation of the terms and conditions of any contract or other agreement to which it may be a party.

8.2 Force Majeure. Neither Party will be held liable or responsible to the other Party nor be deemed to have defaulted under or breached this Agreement for failure or delay in fulfilling or performing any term of this Agreement when such failure or delay is caused by or results from causes beyond the reasonable control of the affected Party (which shall include taking reasonable precautions) including fires, floods, earthquakes, shortages, epidemics, quarantines, embargoes, acts of terrorism, war, acts of war (whether war be declared or not), insurrections, strikes, lockouts, or other labor disturbances, riots, civil commotions, acts of God or acts, omissions, or delays in acting by any Governmental Authority (each, a "**Force Majeure**"). The affected Party will notify the other Party of such Force Majeure as soon as reasonably practical after such occurrence by giving written notice 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 effective only to the extent of and no longer in duration than the Force Majeure causing the failure or delay in performance and the non-performing Party will use every reasonable effort to remedy its inability to perform due to such Force Majeure.

8.3 Governing Law. This Agreement shall be governed by and construed in accordance with the laws of the State of New York, without regard to the conflict of laws principles thereof that would require the application of the law of any other jurisdiction. Except for Financial Disputes which are governed by Section 17.1.2 of the LA, the Parties irrevocably and unconditionally submit to the exclusive jurisdiction of the United States District Court for the Southern District of New York solely and specifically for the purposes of any action or proceeding arising out of or in connection with this Agreement.

8.4 Relationship of the Parties. Each Party shall bear its own costs incurred in the performance of its obligations hereunder without charge or expense to the other Party except as expressly provided in this Agreement. Neither Kiniksa nor Regeneron shall have any responsibility for the hiring, termination or compensation of the other Party's employees or for any employee compensation or benefits of the other Party's employees. No employee or representative of a Party shall have any authority to bind or obligate the other Party to this Agreement for any sum or in any manner whatsoever, or to create or impose any contractual or other liability on the other Party without said Party's approval. For all purposes, and notwithstanding any other provision of this Agreement to the contrary, Regeneron's legal relationship under this Agreement to Kiniksa, and Kiniksa's legal relationship under this Agreement to Regeneron, shall be that of an independent contractor. Nothing in this Agreement shall be construed to establish an employment, agency, joint venture, or partnership

between the Parties or any of their respective Affiliates. For purposes of this Agreement, as of the Effective Date, neither Kiniksa nor any of its Affiliates is an Affiliate of Regeneron or any of its Affiliates, and neither Regeneron nor any of its Affiliates is an Affiliate of Kiniksa or any of its Affiliates

8.5 Assignment. Neither Party may, without the prior written consent of the other Party, delegate, transfer, convey, assign or pledge any of its rights or obligations under this Agreement to any other person, firm or corporation; provided however, that either Party may assign this Agreement in whole or in part to an Affiliate, even without an assignment of the LA. Each Party may assign this Agreement without consent of the other Party to a Third Party in connection with an assignment of the LA. This Agreement shall be binding upon and inure to the benefit of the Parties and their respective successors and permitted assigns.

8.6 Notice. Notice shall be in accordance with Section 17.3 of the LA.

8.7 Compliance with Laws. In the performance of this Agreement, both Parties agree to comply with all applicable Laws.

8.8 Severability. In the event any one or more of the provisions contained in this Agreement should be held invalid, illegal or unenforceable in any respect, the validity, legality and enforceability of the remaining provisions contained herein will not in any way be affected or impaired thereby, unless the absence of the invalidated provision(s) frustrates the purpose of this Agreement (in which case the parties will attempt to replace such invalidated provision with an enforceable provision that most clearly implements such purpose). The Parties will in such an instance use their best efforts to replace the invalid, illegal or unenforceable provision(s) with valid, legal and enforceable provision(s) that, insofar as practical, implement the purposes of this Agreement.

8.9 Accrued Rights. Neither Kiniksa nor Regeneron shall be relieved of any obligations (including payment obligations) of such Party arising prior to termination of this Agreement, including the payment of any non-cancelable costs and expenses (even if such costs and expenses arise following termination or expiration, as the case may be).

8.10 Waiver. The waiver by either Party hereto of any right hereunder or the failure to perform or of a breach by the other Party will not be deemed a waiver of any other right hereunder or of any other breach or failure by said other Party whether of a similar nature or otherwise.

8.11 Headings. Headings in this Agreement are included for ease of reference only and have no legal effect.

8.12 Counterparts. This Agreement may be executed in identical duplicate copies exchanged by facsimile or e-mail (in PDF format) transmission. Each identical counterpart will be deemed an original, but all of which together will constitute one and the same instrument.

IN WITNESS WHEREOF, the Parties have caused this Agreement to be duly executed in their respective names and on their behalf, as of the date first above written.

REGENERON PHARMACEUTICALS, INC.		KINIKSA PHARMACEUTICALS, LTD.	
By:	/s/ Paul Hainsworth	By:	/s/ Thomas Beetham
Name:	Paul Hainsworth	Name:	Thomas Beetham
Title:	VP Supply Chain	Title:	Executive Vice President

Exhibit 21.1

SUBSIDIARIES OF KINIKSA PHARMACEUTICALS, LTD.

Legal Name of Subsidiary	Jurisdiction of Organization
Kiniksa Pharmaceuticals Corp.	Delaware