Table of Contents

UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

FORM 10-Q

(Mark One)

☒ **QUARTERLY REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934**

For the quarterly period ended March 31, 2021
OR

☐ **TRANSITION REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934**

For the transition period from to
Commission file number: 001-38492

Kiniksa Pharmaceuticals, Ltd.

(Exact Name of Registrant as Specified in Its Charter)

Bermuda	**98-1327726**
(State or Other Jurisdiction of Incorporation or Organization)	(I.R.S. Employer Identification No.)

Kiniksa Pharmaceuticals, Ltd.
Clarendon House
2 Church Street
Hamilton HM11, Bermuda
(808) 451-3453
(Address, zip code and telephone number, including area code of principal executive offices)

Kiniksa Pharmaceuticals Corp.
100 Hayden Avenue
Lexington, MA, 02421
(781) 431-9100
(Address, zip code and telephone number, including area code of agent for service)
N/A
(Former name, former address and former fiscal year, if changed since last report)

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	Trading Symbol(s)	Name of each exchange on which registered
Class A Common Shares	KNSA	The Nasdaq Global Select Market

Indicate by check mark whether the registrant (1) has filed all reports required to be filed by Section 13 or 15(d) of the Securities Exchange Act of 1934 during the preceding 12 months (or for such shorter period that the registrant was required to file such reports), and (2) has been subject to such filing requirements for the past 90 days. Yes ☒ No ☐

Indicate by check mark whether the registrant has submitted electronically every Interactive Data File required to be submitted pursuant to Rule 405 of Regulation S-T (§232.405 of this chapter) during the preceding 12 months (or for such shorter period that the registrant was required to submit such files). Yes ☒ No ☐

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

Large Accelerated Filer	☒	Accelerated Filer	☐
Non-accelerated Filer	☐	Smaller Reporting Company	☐
		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 13(a) of the Exchange Act. ☐

Indicate by check mark whether the registrant is a shell company (as defined in Rule 12b-2 of the Exchange Act). Yes ☐ No ☒

As of April 30, 2021, there were 68,332,034 common shares outstanding in aggregate, comprised of:
32,322,276 Class A common shares, par value $0.000273235 per share
1,927,614 Class B common shares, par value $0.000273235 per share
18,024,526 Class A1 common shares, par value $0.000273235 per share
16,057,618 Class B1 common shares, par value $0.000273235 per share

Kiniksa Pharmaceuticals, Ltd.

FORM 10-Q

FOR THE THREE MONTHS ENDED MARCH 31, 2021

TABLE OF CONTENTS

SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This Quarterly Report on Form 10-Q, or Quarterly Report, contains forward-looking statements. All statements other than statements of historical facts contained in this Quarterly Report including statements regarding our future results of operations and financial position, expected timeline for our cash, cash equivalents and short-term investments, business strategy, product development, prospective products and product candidates, their expected properties, performance, market opportunity and competition, drug product supply, collaborators, license and other strategic arrangements, the expected timeline for achievement of our clinical milestones, the timing of, and potential results from, clinical and other trials, potential marketing authorization from the FDA or regulatory authorities in other jurisdictions, potential coverage and reimbursement for our products and product candidates, if approved, commercial strategy and pre-commercial activities, research and development costs, timing of regulatory filings and feedback, timing and likelihood of success, plans and objectives of management for future operations and funding requirements 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,” “goal,” “design,” “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 Quarterly Report 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 Quarterly Report and are subject to a number of risks, uncertainties and assumptions described under the sections in this Quarterly Report entitled “Risk Factors” and “Management’s Discussion and Analysis of Financial Condition and Results of Operations” and elsewhere in this Quarterly Report. These forward-looking statements are subject to numerous risks and uncertainties, including, without limitation, the following:

- the impact COVID-19 pandemic on our business, including our clinical trials and operations;
- our status as a commercial-stage biopharmaceutical company and our expectation to incur losses in the future;
- our future capital needs and our need to raise additional funds, including through public or private securities offerings, debt financings or other sources, which may include licensing, collaborations or other strategic transactions or arrangements;
- the lengthy and expensive clinical development process with its uncertain outcome and potential for clinical failure or delay, including due to the COVID-19 pandemic;
- the decision by any applicable regulatory authority whether to clear our current or future 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 undertake business combinations, collaborations or other strategic transactions;
- our ability to have our product candidates manufactured in accordance with regulatory requirements;
- the market acceptance of our product candidates;

- our ability to timely and successfully commercialize ARCALYST (rilonacept) and to develop and commercialize our current and future product candidates, if approved;
- competitive and potentially competitive products and technologies;
- 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 ARCALYST or any of our current or future product candidates or to require extensive 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;
- ownership concentration of our executive officers, certain members of senior management and affiliated shareholders may prevent our shareholders from influencing significant corporate decisions;
- our ability to attract and retain skilled personnel; and
- our ability to execute on our strategy.

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 Quarterly Report 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 Quarterly Report and the documents that we reference in this Quarterly Report 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.

SUMMARY RISK FACTORS

Our business is subject to numerous risks and uncertainties, including those described in Part II, Item 1A. “Risk Factors” in this Quarterly Report. You should carefully consider these risks and uncertainties when investing in our Class A common shares. The principal risks and uncertainties affecting our business include the following:

- the COVID-19 pandemic, and related measures taken in response or the easing of such measures, could have an adverse impact on our business and operations as well as those of our manufacturers, contract research organizations and other third parties with whom we conduct business or otherwise engage, including regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position;

- we have not previously generated any product revenue, have incurred significant operating losses since our inception, and expect to incur significant operating losses for the foreseeable future and may never achieve or maintain profitability;

- we will require significant additional funding to continue commercialization of ARCALYST or any other products, if approved, and the development and commercialization of our product candidates, if approved, and to identify, discover, develop or acquire additional product candidates, and if we are unable to raise capital on acceptable terms when needed, or at all, we could be forced to delay, reduce or cease one or more of our product development plans, research and development programs or other operations or commercialization efforts;

- we depend heavily on the commercial success of ARCALYST, and have limited experience commercializing a therapeutic, supporting sales, marketing, and distribution activities and maintaining applicable infrastructure for these activities either directly and/or through agreements with third parties; as a result we may not be successful in commercializing ARCALYST, or any future approved product candidates, thus potentially impairing the commercial potential of ARCALYST and our other product candidates, if approved, to generate any revenue;

- we depend heavily on the success of one or more of our product candidates, which are in various stages of product developments, and cannot give any assurance that we will be able to advance our product candidates in clinical development, obtain regulatory approval and ultimately commercialize one or more of our product candidates on a timely basis, if at all;

- ARCALYST in recurrent pericarditis, as well as our current or future product candidates, may not gain market acceptance by physicians, patients, or third-party payors, in which case our ability to generate product revenues will be impaired;

- successful commercialization of our products and product candidates, if approved, will depend in part on the extent to which third-party payors provide funding, establish favorable coverage and pricing policies and set adequate reimbursement levels for our products and product candidates, if approved, and failure to obtain or maintain coverage and adequate reimbursement for our products and product candidates, if approved, could limit our ability to market those products and decrease our ability to generate revenue;

- the incidence and prevalence for target patient populations of our products and product candidates have not been established with precision, and if the market opportunities for our products and product candidates, if approved, 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;

- clinical development of our product candidates is a lengthy and expensive process with uncertain timelines, costs and outcomes;

- we may encounter substantial delays in our current or planned pre-clinical and clinical trials, including as a result of delays in obtaining regulatory approvals for indications, site activation, enrollment, and conduct of the trials, which could delay or prevent our product development activities;

- we rely on third parties, including independent contract research organizations, or CROs, to activate our sites, conduct or otherwise support our research activities, preclinical studies and clinical trials for our product candidates, and these third parties may not perform satisfactorily, which could delay, prevent or impair our product development activities;

- we contract with third parties, including independent contract manufacturing organizations, or CMOs, to manufacture our product candidates for preclinical and clinical development and do so for our commercial supply of ARCALYST as well as for supply of drug substance and drug product for our product candidates, and these third parties may not perform satisfactorily, which could delay, prevent or impair our product development activities, regulatory approval, and commercialization efforts for our product candidates;

- all of our products and product candidates have been licensed or acquired from other parties; if we are unable to adequately protect such products and product candidates, or to secure and maintain freedom to operate, others could preclude us from commercializing our products and product candidates, if approved, or compete against us more directly;

- we face significant competition from other biotechnology and pharmaceutical companies, which may result in others discovering, developing or commercializing drugs before or more successfully than us;

- we may not be successful in executing our growth strategy to identify, discover, develop, license or acquire additional product candidates or technologies, and our strategy may not deliver anticipated results or we may refine or otherwise alter our growth strategy;

- we may seek to acquire businesses or undertake business combinations, collaborations or other strategic transactions which may not be successful or on favorable terms, if at all, and we may not realize the intended benefits of such transactions; and

- concentration of ownership of the voting power of our common shares may prevent new investors from influencing significant corporate decisions and may have an adverse effect on the price of our Class A common shares.

INDUSTRY AND OTHER DATA

Unless otherwise indicated, certain industry data and market data included in this Quarterly Report 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 Quarterly Report 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 Quarterly Report is reliable.

ARCALYST is a registered trademark of Regeneron Pharmaceuticals, Inc. Solely for convenience, trademarks, service marks, and trade names referred to in this Quarterly Report may be listed without identifying symbols.

Part I — Financial Information

Item 1. Financial Statements (unaudited)

KINIKSA PHARMACEUTICALS, LTD.
CONSOLIDATED BALANCE SHEETS
(In thousands, except share and per share amounts)
(Unaudited)

	March 31, 2021	December 31, 2020
Assets		
Current assets:		
Cash and cash equivalents	$ 119,856	$ 114,038
Short-term investments	144,169	209,444
Restricted cash	210	210
Inventory	2,189	—
Prepaid expenses and other current assets	9,140	9,557
Total current assets	275,564	333,249
Property and equipment, net	3,815	4,051
Operating lease right-of-use assets	5,982	6,566
Other long-term assets	5,797	5,588
Intangible asset	20,000	—
Deferred tax assets	—	10
Total assets	$ 311,158	$ 349,464
Liabilities and Shareholders' Equity		
Current liabilities:		
Accounts payable	$ 2,763	$ 503
Accrued expenses	30,316	29,199
Operating lease liabilities	2,374	2,107
Other current liabilities	—	37
Total current liabilities	35,453	31,846
Non-current liabilities:		
Non-current operating lease liabilities	4,200	4,878
Other long-term liabilities	809	805
Total liabilities	40,462	37,529
Commitments and contingencies (Note 13)		
Shareholders' equity:		
Class A common shares, par value of $0.000273235 per share; 32,320,276 shares and 31,777,420 shares issued and outstanding as of March 31, 2021 and December 31, 2020, respectively	8	8
Class B common shares, par value of $0.000273235 per share; 1,927,614 shares and 2,355,458 shares issued and outstanding as of March 31, 2021 and December 31, 2020, respectively	1	1
Class A1 common shares, $0.000273235 par value; 18,024,526 issued and outstanding as of March 31, 2021 and December 31, 2020	5	5
Class B1 common shares, $0.000273235 par value; 16,057,618 shares issued and outstanding as of March 31, 2021 and December 31, 2020	4	4
Additional paid-in capital	837,656	829,424
Accumulated other comprehensive loss	(21)	(34)
Accumulated deficit	(566,957)	(517,473)
Total shareholders' equity	270,696	311,935
Total liabilities and shareholders' equity	$ 311,158	$ 349,464

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)
(Unaudited)

	Three Months Ended March 31,	
	2021	2020
Operating expenses:		
Research and development	$ 28,683	$ 20,901
Selling, general and administrative	20,600	8,486
Total operating expenses	49,283	29,387
Loss from operations	(49,283)	(29,387)
Interest income	9	789
Loss before (provision) benefit for income taxes	(49,274)	(28,598)
(Provision) benefit for income taxes	(210)	2,179
Net loss	$ (49,484)	$ (26,419)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.72)	$ (0.48)
Weighted average common shares outstanding—basic and diluted	68,269,486	55,322,690
Comprehensive loss:		
Net loss	$ (49,484)	$ (26,419)
Other comprehensive income (loss):		
Unrealized gain on short-term investments and currency translation adjustments, net of tax	13	207
Total other comprehensive income	13	207
Total comprehensive loss	$ (49,471)	$ (26,212)

The accompanying notes are an integral part of these consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.
CONSOLIDATED STATEMENTS OF SHAREHOLDERS' EQUITY
(In thousands, except share amounts)
(Unaudited)

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income (Loss)	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
Balances at December 31, 2020	68,215,022	$ 18	$ 829,424	$ (34)	$ (517,473)	$ 311,935
Issuance of Class A common shares under incentive award plans	115,012	—	1,106	—	—	1,106
Share-based compensation expense	—	—	7,126	—	—	7,126
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	13	—	13
Net loss	—	—	—	—	(49,484)	(49,484)
Balances at March 31, 2021	68,330,034	$ 18	$ 837,656	$ (21)	$ (566,957)	$ 270,696

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
Balances at December 31, 2019	54,937,628	$ 15	$ 581,467	$ 33	$ (356,092)	$ 225,423
Issuance of Class A common shares under incentive award plans	643,867	—	2,414	—	—	2,414
Share-based compensation expense	—	—	4,209	—	—	4,209
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	207	—	207
Net loss	—	—	—	—	(26,419)	(26,419)
Balances at March 31, 2020	55,581,495	$ 15	$ 588,090	$ 240	$ (382,511)	$ 205,834

The accompanying notes are an integral part of these consolidated financial statements.

KINIKSA PHARMACEUTICALS, LTD.
CONSOLIDATED STATEMENTS OF CASH FLOWS
(In thousands)
(Unaudited)

	Three Months Ended March 31,	
	2021	**2020**
Cash flows from operating activities:		
Net loss	$ (49,484)	$ (26,419)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation and amortization expense	312	594
Share-based compensation expense	7,126	4,209
Non-cash lease expense	593	300
Amortization of premiums and accretion of discounts on short-term investments	410	(228)
Deferred income taxes	10	(1,038)
Changes in operating assets and liabilities:		
Prepaid expenses and other assets	408	(598)
Other long-term assets	(231)	—
Accounts payable	72	(1,197)
Accrued expenses and other liabilities	1,079	(7,061)
Operating lease liabilities	(421)	(413)
Other long-term liabilities	4	55
Net cash used in operating activities	(40,122)	(31,796)
Cash flows from investing activities:		
Purchases of property and equipment	(54)	(223)
Purchases of short-term investments	(74,362)	(26,187)
Proceeds from the maturities of short-term investments	139,250	115,200
Intangible asset acquired	(20,000)	—
Net cash provided by investing activities	44,834	88,790
Cash flows from financing activities:		
Proceeds from issuance of Class A common shares under incentive award plans	1,106	2,414
Net cash provided by financing activities	1,106	2,414
Net increase in cash, cash equivalents and restricted cash	5,818	59,408
Cash, cash equivalents and restricted cash at beginning of period	114,248	47,138
Cash, cash equivalents and restricted cash at end of period	$ 120,066	$ 106,546

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)
(Unaudited)

1. Nature of the Business and Basis of Presentation

Kiniksa Pharmaceuticals, Ltd. (the "Company") is a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. The Company was incorporated in July 2015 as a Bermuda exempted company. The Company's portfolio of assets is based on strong biologic rationale or validated mechanisms, target underserved conditions and offer the potential for differentiation.

The Company is subject to risks and uncertainties common to early, commercial-stage companies in the biopharmaceutical industry and global health, societal, economic and market conditions, including from the impact from the coronavirus disease 2019 ("COVID-19") pandemic. While the U.S. Food and Drug Administration (the "FDA"), approved the supplemental Biologics License Application ("sBLA") for ARCALYST® (rilonacept) for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021, the Company has limited experience obtaining regulatory approvals. There can be no assurance that the Company's research and development of its current or future product candidates will be successfully completed, that adequate protection for the Company's technology will be obtained or maintained, that any current or future product candidates will obtain necessary government regulatory approval or that ARCALYST or any of the Company's current or future product candidates, if approved, will be commercially viable. Upon approval from the FDA of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence, the Company assumed the sales and distribution of ARCALYST for the previously approved indications for the treatment of Cryopyrin-Associated Periodic Syndromes ("CAPS"), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and for the maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist ("DIRA") in adults and children 12 years of age and older in the United States and will evenly split profits on sales with Regeneron, Inc. ("Regeneron"), after deducting certain commercialization expenses subject to specified limits. The Company has limited experience conducting sales and marketing activities for therapeutic products, and as a result it may never be able to successfully commercialize ARCALYST or any other marketable product in the future. Furthermore, the Company operates in an environment of rapid technological innovation and substantial competition from pharmaceutical and biotechnology companies. In addition, the Company is dependent upon the services of its employees, consultants and other third parties, including contract research organizations ("CROs"), and contract manufacturing organizations ("CMOs") for the development and manufacture of its product candidates, respectively. Even if the Company's current and future product development efforts are successful, it is uncertain when, if ever, the Company will realize significant revenue from product sales or be profitable.

Risk and Uncertainties Related to COVID-19

In addition to risks and uncertainties common to early, commercial-stage companies in the Company's industry, the Company is subject to global societal, healthcare, economic and market conditions, including from the impact of the COVID-19 pandemic and measures taken in response to the pandemic or the easing of such measures, which continue to evolve. In December 2019, COVID-19 surfaced in Wuhan, China. The virus spread globally and was declared a pandemic by the World Health Organization. The impact of this pandemic has been and will likely continue to be extensive on many aspects of society, which has resulted in and will likely continue to result in significant disruptions to healthcare systems, the global economy, as well as businesses and capital markets around the world.

In an effort to halt the spread of the COVID-19 pandemic, federal and state governments in the United States and the governments of other countries around the globe have implemented various measures in response to the pandemic, including significant restrictions on businesses and travel as well as social-distancing measures and the easing of such measures. In response to the COVID-19 pandemic and measures introduced by federal and state governments in the United States, the Company implemented workplace protocols in the jurisdictions where it has facilities. While the majority of the Company's employees are able to carry out their responsibilities working outside of the Company's physical locations, for the Company's essential workers and those choosing to return to the Company's offices to carry

out their responsibilities, the Company implemented additional safety measures, including occupancy limits, restricting business travel, providing and requiring the use of personal protective equipment, self-screening prior to accessing the Company's facilities, among other things. As these measures implemented by federal and state governments in the United States in response to the pandemic continue to evolve, the Company continues to monitor the developments, restrictions and requirements in jurisdictions where it has offices, and plans to update the protocols for its offices as applicable.

The COVID-19 pandemic, and measures undertaken in response to the pandemic, or the easing of any of such measures, may cause significant disruptions in the Company's business or operations as well as in the business and operations of the Company's CMOs, CROs and other third parties with whom the Company conducts business or otherwise engages now or in the future, including as a result of staffing shortages or reprioritizations, production slowdowns or stoppages, and disruptions in delivery systems. The COVID-19 pandemic may also adversely impact the Company's preclinical studies and clinical trials, which could impede, delay, limit or prevent the clinical development of the Company's product candidates, lead to the delay or denial of regulatory approval of its product candidates or delay or adversely impact the Company's commercialization activities, which would materially adversely affect the Company's business and operations, including its ability to generate revenue. Moreover, the COVID-19 pandemic is still impacting the global economy, including the U.S. economy, with the potential for the economic downturn to be severe and prolonged. A severe or prolonged economic downturn could result in continued disruptions in the financial markets, which could adversely impact the Company's ability to raise additional capital when needed or on acceptable terms, if at all.

While the Company continuously looks to identify business-critical activities and to develop contingencies and mitigation strategies for those activities to potentially minimize the impact of the COVID-19 pandemic on its business and operations, there can be no assurance that it will be able to identify all such activities or that any identified contingencies and mitigation strategies will be effective. Further, the COVID-19 pandemic, and measures undertaken in response to the pandemic continue to rapidly evolve, including the presence of new variants. There is uncertainty relating to the potential effect of the pandemic on the Company's business and operations. The extent of the impact on the Company's business and operations will depend on future developments, which are highly uncertain and cannot be predicted with confidence, such as the ultimate spread of the disease, duration of the pandemic, business and travel restrictions and social distancing measures, and the effectiveness of these and other actions taken to contain, prevent and treat the disease as well as the impact of the easement of any such restrictions, measures and actions.

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 subsidiaries, Kiniksa Pharmaceuticals Corp. ("Kiniksa US"), Primatope Therapeutics, Inc. ("Primatope") and Kiniksa Pharmaceuticals (UK), Ltd. ("Kiniksa UK") as well as the subsidiaries of Kiniksa UK, Kiniksa Pharmaceuticals (Germany) GmbH ("Kiniksa Germany"), Kiniksa Pharmaceuticals (France) SARL ("Kiniksa France"), and Kiniksa Pharmaceuticals GmbH ("Kiniksa Switzerland"), after elimination of all significant intercompany accounts and transactions.

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 share-based awards. Estimates are periodically reviewed in light of changes in circumstances, facts and experience.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

Changes in estimates are recorded in the period in which they become known. Actual results could differ from those estimates.

Reporting and Functional Currency

The financial results of the Company's global activities are reported in U.S. dollars (“USD”) and its foreign subsidiaries either utilize USD or their respective local currency to be their functional currency.

Transactions in other currencies are recorded in the functional currency at the rate of exchange prevailing when the transactions occur. Monetary assets and liabilities denominated in other currencies are re-measured into the functional currency at the rate of exchange in effect at the balance sheet date. Exchange rate gains and losses arising from re-measurement of foreign currency-denominated monetary assets and liabilities are included in income or losses in the period in which they occur.

For the Company’s foreign subsidiaries where the local currency is the functional currency, assets and liabilities denominated in local currencies are translated into USD at end-of-period exchange rates and the resulting translation adjustments are reported as a component of accumulated other comprehensive gain (loss) within shareholders' equity (deficit).

Unaudited Interim Consolidated Financial Information

The accompanying unaudited consolidated financial statements have been prepared in accordance with GAAP for interim financial information. The accompanying unaudited consolidated financial statements do not include all of the information and footnotes required by GAAP for complete consolidated financial statements. The accompanying year-end consolidated balance sheet was derived from audited financial statements but does not include all disclosures required by GAAP. The unaudited interim consolidated financial statements have been prepared on the same basis as the audited annual consolidated financial statements and, in the opinion of management, reflect all adjustments, which include only normal recurring adjustments, necessary for the fair statement of the Company’s financial position as of March 31, 2021 and the results of its operations for the three months ended March 31, 2021 and 2020 and its cash flows for the three months ended March 31, 2021 and 2020. The results for the three months ended March 31, 2021 are not necessarily indicative of results to be expected for the year ending December 31, 2021, any other interim periods or any future year or period.

Liquidity

In accordance with Accounting Standards Update (“ASU”) No. 2014-15, Disclosure of Uncertainties about an Entity’s Ability to Continue as a Going Concern (Subtopic 205-40), the Company has evaluated whether there are conditions and events, considered in the aggregate, that raise substantial doubt about the Company’s ability to continue as a going concern within one year after the date the consolidated financial statements are issued. As of March 31, 2021, the Company had an accumulated deficit of $566,957. During the three months ended March 31, 2021, the Company incurred a net loss of $49,484, used $40,122 of cash in operating activities, and paid $20,000 upon the achievement of specified regulatory milestone (see Note 10). The Company expects to continue to generate operating losses for the foreseeable future. As of March 31, 2021, the Company had cash, cash equivalents and short-term investments of $264,025.

Based on its current operating plan, the Company expects that its cash, cash equivalents and short-term investments will be sufficient to fund its operations and capital expenditure requirements for at least twelve months from the issuance date of these consolidated financial statements. The future viability of the Company beyond that point is dependent on its ability to raise additional capital to finance its operations. The Company will need to finance its operations through public or private securities offerings, debt financings, government funding or grants, or other sources, which may include licensing, collaborations or other strategic transactions or arrangements. Although the Company has

been successful in raising capital in the past, there is no assurance that it will be successful in obtaining such additional financing on terms acceptable to the Company, if at all. If the Company is unable to obtain funding, the Company could be forced to delay, reduce or eliminate some or all of its commercialization efforts, research and development programs for product candidates or product portfolio expansion, which could adversely affect its business prospects, or the Company may be unable to continue operations.

2. Summary of Significant Accounting Policies

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 March 31, 2021 and December 31, 2020, cash and cash equivalents consisted principally of U.S. Treasury notes, amounts held in money market accounts and cash on deposit at commercial banks.

Short-Term Investments

The Company generally invests its excess cash in money market funds and short-term investments in U.S. Treasury notes. Such investments which are included in short-term investments on the Company's consolidated balance sheets are considered available-for-sale debt securities and are reported at fair value with unrealized gains and losses included as a component of shareholders’ equity. Realized gains and losses, if any, on short-term investments are included in interest income.

If the estimated fair value of a debt security is below its carrying value, the Company evaluates whether it is more likely than not that we will sell the security before its anticipated recovery in market value and whether evidence indicating that the cost of the investment is recoverable within a reasonable period of time outweighs evidence to the contrary. The Company also evaluates whether or not it intends to sell the investment. If the impairment is considered to be other-than-temporary, the security is written down to its estimated fair value. In addition, we consider whether credit losses exist for any securities. A credit loss exists if the present value of cash flows expected to be collected is less than the amortized cost basis of the security. Other-than-temporary declines in estimated fair value and credit losses are included in investment and other income, net.

Concentrations of Credit Risk

Financial instruments that potentially subject the Company to concentrations of credit risk consist principally of cash, cash equivalents and short-term investments. At March 31, 2021 and December 31, 2020, substantially all of the Company’s cash, cash equivalents and short-term investments 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, cash equivalents and short-term investments and does not believe that it is subject to unusual credit risk beyond the normal credit risk associated with commercial banking relationships.

Restricted Cash

In conjunction with the Company’s lease agreement entered into in March 2018, the Company maintains a letter of credit for the benefit of the landlord. As of March 31, 2021 and December 31, 2020 the underlying cash balance of $210 securing this letter of credit, was classified as current in its consolidated balance sheet.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

Fair Value Measurements

Certain assets and liabilities of the Company are carried at fair value under GAAP. Fair value is defined as the exchange price that would be received for an asset or paid to transfer a liability (an exit price) in the principal or most advantageous market for the asset or liability in an orderly transaction between market participants on the measurement date. Valuation techniques used to measure fair value must maximize the use of observable inputs and minimize the use of unobservable inputs. Financial assets and liabilities carried at fair value are to be classified and disclosed in one of the following three levels of the fair value hierarchy, of which the first two are considered observable and the last is considered unobservable:

- Level 1—Quoted prices in active markets for identical assets or liabilities.
- Level 2—Observable inputs (other than Level 1 quoted prices), such as quoted prices in active markets for similar assets or liabilities, quoted prices in markets that are not active for identical or similar assets or liabilities, or other inputs that are observable or can be corroborated by observable market data.
- Level 3—Unobservable inputs that are supported by little or no market activity that are significant to determining the fair value of the assets or liabilities, including pricing models, discounted cash flow methodologies and similar techniques.

The Company's restricted cash, which is held in a money market fund, is carried at fair value, determined based on Level 1 inputs in the fair value hierarchy described above (see Note 3). The Company's cash equivalents and short-term investments, 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.

Leases

At the inception of an arrangement, the Company determines whether the arrangement is or contains a "lease" as defined by ASU No. 2016-02, *Leases (Topic 842)* ("ASC 842"). A lease is an arrangement, or part of an arrangement, that conveys the right to control the use of identified property, plant, or equipment (an identified asset) for a period of time in exchange for consideration. The Company determines if the arrangement conveys the right to control the use of an identified asset for a period of time. It assesses throughout the period of use whether the Company has both of the following (1) the right to obtain substantially all of the economic benefits from use of the identified asset and (2) the right to direct the use of the identified asset. This determination is reassessed if the terms of the arrangement are changed. Leases are classified as operating or finance leases based on the terms of the lease agreement and certain characteristics of the identified asset. Right-of-use ("ROU") assets and lease liabilities are recognized at lease commencement date based on the present value of the minimum future lease payments.

Most leases with a term greater than one year are recognized on the balance sheet as ROU assets with corresponding lease liabilities and, if applicable, long-term lease liabilities. The Company has elected not to recognize leases with a term of one year or less on its balance sheet. Operating leases, ROU assets and their corresponding lease liabilities are recorded based on the present value of lease payments over the expected remaining lease term. However, certain adjustments to the ROU assets may be required for items such as incentives received. The interest rate implicit in lease arrangements is typically not readily determinable. As a result, the Company utilizes its incremental borrowing rate, which is the rate incurred to borrow on a collateralized basis over a similar term an amount equal to the lease payments in a similar economic environment.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

In accordance with the guidance in ASC 842, components of a lease should be split into three categories: lease components (e.g., land, building, etc.), non-lease components (e.g., common area maintenance, consumables, etc.), and non-components (e.g., property taxes, insurance, etc.); then the fixed and in-substance fixed contract consideration (including any related to non-components) must be allocated based on fair values to the lease components and non-lease components.

The Company has elected to account for the lease and non-lease components of each of its operating leases as a single lease component and allocate all of the arrangement consideration to the lease component only. The lease component results in an operating ROU asset being recorded on the balance sheet and amortized on a straight-line basis as lease expense.

Inventory

Inventories are stated at the lower of cost or estimated net realizable value with cost based on the first-in first-out method. Inventory that can be used in either the production of clinical or commercial products is expensed as research and development costs when identified and labeled for use in clinical trials as the products are required to be re-labeled for alternative uses. Prior to the regulatory approval of its drug candidates, the Company incurs expenses for the manufacture of product candidate supplies to support clinical development that could potentially be available to support the commercial launch of those therapeutics. Until the date at which regulatory approval has been received or is otherwise considered probable, the Company records all such costs as research and development expenses. The Company performs an assessment of the recoverability of capitalized inventories during each reporting period and writes down any excess and obsolete inventory to its net realizable value in the period in which the impairment is first identified. Such impairment charges, should they occur, are recorded as a component of cost of sales in the Company's consolidated statements of operations and comprehensive loss. The determination of whether inventory costs will be realizable requires the use of estimates by management. If actual market conditions are less favorable than projected by management, additional write-downs of inventory may be required.

Intangible Assets

Upon FDA approval and commercial launch of ARCALYST in March 2021, the Company capitalized the $20,000 milestone payment to Regeneron for a specified regulatory milestone as a finite-lived intangible asset. The intangible asset will be amortized on a straight-line basis over the life of the underlying intellectual property of 20 years. Amortization expense will be recorded as cost of goods sold in the Company’s consolidated statement of operations and comprehensive loss.

Impairment of long-lived assets

The Company assesses the impairment of long-lived assets, including intangible assets and property and equipment, whenever events or changes in business circumstances indicate that the carrying amounts of the assets may not be fully recoverable. When such events occur, the Company determines whether there has been an impairment in value by comparing the asset’s carrying value with its fair value, as measured by the anticipated undiscounted net cash flows of the asset. If an impairment in value exists, the asset is written down to its estimated fair value. The Company has not recognized any impairment losses through March 31, 2021 and there have been no events that triggered an impairment analysis.

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 preclinical and clinical development activities and clinical trials as well as to manufacture

clinical trial materials. Non-refundable prepayments determined to be used within one year for goods or services that will be used or rendered for future research and development activities are recorded as prepaid expenses. Non-refundable prepayments or minimum balance requirements associated to clinical trials determined to not be used within one year are classified as other long term assets. 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. Milestone and other payments made to third parties with respect to in-process research and development, in accordance with the Company's license, acquisition and other similar agreements are expensed when determined to be probable and estimable.

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 and development costs. When billing terms under these contracts do not coincide with the timing of when the work is performed, the Company is required to make estimates of outstanding obligations to those third parties as of the end of the reporting period. Any accrual estimates are based on a number of factors, including the Company's knowledge of the progress towards completion of the research and development activities, invoicing to date under the contracts, communication from the research institution or other companies of any actual costs incurred during the period that have not yet been invoiced, and the costs included in the contracts. 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.

Share-Based Compensation

The Company measures all share-based awards granted to employees and directors based on their fair value on the date of grant. The Company issues share-based awards with both service-based vesting conditions and performance-based vesting conditions. The Company recognizes compensation expense for awards with service conditions on a straight-line basis over the requisite service period. For awards with performance conditions, the Company recognizes compensation expense when the achievement of the performance milestone is probable and estimable through the vest date.

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.

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 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 9). Prior to May 2018, the Company was a private company and, accordingly, lacks company-specific historical and implied volatility information for its shares. Therefore, it estimates its expected share price volatility based on a blend of the historical volatility of the Company's and publicly traded peer companies share price 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

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

Company has never paid cash dividends on common shares and does not expect to pay any cash dividends in the foreseeable future.

The fair value of each restricted share unit award is based on the closing price of the Company's Class A common shares on the date of grant. Restricted share unit awards with an associated performance condition are evaluated on a regular basis for probability of achievement to determine the timing of recording share-based compensation expense in the Company's consolidated statements of operations and comprehensive loss.

Comprehensive Loss

Comprehensive loss includes net loss as well as other changes in shareholders' equity that result from transactions and economic events other than those with shareholders. For the three months ended March 31, 2021 and 2020, the Company's other comprehensive loss was comprised of unrealized gain (loss) on short-term investments as well as cumulative translation adjustments, net of tax.

Net Loss per Share

The Company follows the two-class method when computing net loss per share as the Company has issued shares that meet the definition of participating securities. The two-class method determines net loss per share for each class of common shares 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 loss per share attributable to common shareholders is computed by dividing the net loss attributable to common shareholders by the weighted average number of common shares outstanding for the period. Diluted net loss attributable to common shareholders is computed by adjusting net loss attributable to common shareholders to reallocate undistributed earnings based on the potential impact of dilutive securities. Diluted net loss per share attributable to common shareholders is computed by dividing the diluted net 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 and unvested restricted common shares are considered potential dilutive common shares.

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 three months ended March 31, 2021 and 2020.

Income Taxes

As an exempted company incorporated under the laws of 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 losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards will be available to the Company for those losses. The Company's wholly owned U.S. subsidiaries, Kiniksa US and Primatope, are subject to federal and state income taxes in the United States. The Company's wholly owned subsidiary Kiniksa UK, and its wholly owned subsidiaries, Kiniksa Germany, Kiniksa France, and Kiniksa Switzerland, are subject to taxation in their respective countries. Certain of the Company's subsidiaries, primarily Kiniksa US and Kiniksa Germany, operate under cost-plus arrangements.

The Company's U.S. provision for income taxes relates to current tax expense associated with the taxable income in the United States of its wholly owned subsidiary, Kiniksa US. The current income tax expense is a result of

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

the taxable income earned by Kiniksa US under its cost-plus arrangement offset in part by tax benefits from the U.S. federal and state research and development credits, the Foreign Derived Intangible Income ("FDII") deduction and share-based compensation taxable events. The Company has recorded an immaterial foreign provision for income taxes related to income in non-U.S. subsidiaries.

The Company provides for income taxes on an interim basis according to management's estimate of the effective tax rate expected to be applicable for the full fiscal year. Subsidiaries with losses for which no benefit can be claimed are excluded from this calculation, and their tax is recorded discretely in the period it arises. Certain other items such as changes in tax rates, tax benefits or expense related to settlements of share-based payment awards, changes in the valuation allowance against deferred tax, and uncertain tax positions are treated as discrete items and are recorded in the period in which they arise.

Recently Adopted Accounting Pronouncements

In December 2019, the FASB issued ASU 2019-12, *Simplifying the Accounting for Income Taxes (Topic 740) ("ASU 2019-12")*. The objective of the standard is to improve areas of GAAP by removing certain exceptions permitted by ASC Topic 740 Income Taxes and clarifying existing guidance to facilitate consistent application. The Company adopted the standard on January 1, 2021. The adoption of ASU 2019-12 did not have a material impact on the Company's consolidated financial statements and related disclosures.

3. Fair Value of Financial Assets and Liabilities

The following tables present information about the Company's financial instruments 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 March 31, 2021 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 210	$ —	$ —	$ 210
Cash equivalents — money market funds	33,833	—	—	33,833
Cash equivalents — U.S. Treasury notes	—	65,099	—	65,099
Short-term investments — U.S. Treasury notes	—	144,169	—	144,169
	$ 34,043	$ 209,268	$ —	$ 243,311

	Fair Value Measurements as of December 31, 2020 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 210	$ —	$ —	$ 210
Cash equivalents — money market funds	22,942	—	—	22,942
Cash equivalents — U.S. Treasury notes	—	72,695	—	72,695
Short-term investments — U.S. Treasury notes	—	209,444	—	209,444
	$ 23,152	$ 282,139	$ —	$ 305,291

During the three months ended March 31, 2021 and the year ended December 31, 2020 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 and short-term investments as of March 31, 2021 and December 31, 2020 included U.S. Treasury notes, which are not traded on a daily basis and, therefore, represent a Level 2 measurement in the fair value hierarchy at each period end.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Fair Value
March 31, 2021				
Cash equivalents — U.S. Treasury notes	$ 65,099	$ —	$ —	$ 65,099
Short-term investments — U.S. Treasury notes	144,160	9	—	144,169
	$ 209,259	$ 9	$ —	$ 209,268

	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Fair Value
December 31, 2020				
Cash equivalents — U.S. Treasury notes	$ 72,694	$ 1	$ —	$ 72,695
Short-term investments — U.S. Treasury notes	209,459	4	(19)	209,444
	$ 282,153	$ 5	$ (19)	$ 282,139

As of March 31, 2021, the Company held seven securities that were in an unrealized loss position. The aggregate fair value of securities held by the Company in an unrealized loss position was $28,999 at March 31, 2021. As of December 31, 2020, the Company held 17 securities that were in an unrealized loss position. The aggregate fair value of securities held by the Company in an unrealized loss position was $107,753 at December 31, 2020. As of both March 31, 2021 and December 31, 2020, these securities were held by the Company in an unrealized loss position for less than 12 months. The Company determined that there was no material change in the credit risk of these securities. As a result, the Company determined it did not hold any investments with an other-than-temporary impairment as of March 31, 2021 and December 31, 2020.

4. Inventory

During the three months ended March 31, 2021, the Company commenced the capitalization of ARCALYST inventory in connection with receiving FDA approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older. As of March 31, 2021, the Company identified and relabeled ARCALYST product previously acquired initially intended to support the clinical trials as inventory to be sold as commercial product classified as finished goods. The value of this inventory had previously been expensed to research and development costs and is currently carried at zero cost. As of December 31, 2020, the Company did not have inventory.

Inventory consisted of the following:

	March 31, 2021
Raw materials	$ —
Work-in-process	—
Finished Goods	2,189
	$ 2,189

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

5. Property and Equipment, Net

Property and equipment, net consisted of the following:

	March 31, 2021	December 31, 2020
Furniture, fixtures and vehicles	$ 62	$ 62
Computer hardware and software	349	349
Leasehold improvements	3,667	3,667
Lab equipment	4,602	4,602
Construction in progress	155	101
Total property and equipment	8,835	8,781
Less: Accumulated depreciation	(5,020)	(4,730)
Total property and equipment, net	$ 3,815	$ 4,051

Depreciation expense was $290 and $594 during the three months ended March 31, 2021 and 2020, respectively.

6. Intangible Assets

Intangible assets, net of accumulated amortization, impairment charges and adjustments are summarized in the following table. As of December 31, 2020, the Company did not have intangible assets.

		As of March 31, 2021		
	Estimated life	Cost	Accumulated Amortization	Net
Regulatory milestone	20 years	$ 20,000	$ -	$ 20,000
		$ 20,000	$ -	$ 20,000

As of March 31, 2021	
2021 (remaining nine months)	$ 750
2022	1,000
2023	1,000
2024	1,000
2025	1,000
2026	1,000
2027 and thereafter	14,250

7. Accrued Expenses

Accrued expenses consisted of the following:

	March 31, 2021	December 31, 2020
Accrued research and development expenses	$ 20,038	$ 16,945
Accrued employee compensation and benefits	7,130	7,704
Accrued legal and professional fees	2,857	3,988
Other	291	562
	$ 30,316	$ 29,199

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

8. Common Shares

On May 18, 2020, the Company completed a follow-on offering of 2,760,000 Class A common shares, inclusive of the exercise of the underwriters' overallotment option at a public offering price of $18.25 and a concurrent private placement of 1,600,000 Class A1 common shares at an offering price of $18.25 per share for aggregate gross proceeds of $79,570. The aggregate net proceeds to the Company from the follow-on offering and concurrent private placement, inclusive of the over-allotment option exercise, was $74,495 after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

On July 24, 2020, the Company completed a follow-on offering of 5,952,381 Class A common shares, at a public offering price of $21.00 and a concurrent private placement of 1,428,572 Class A1 common shares at an offering price of $21.00 per share for aggregate gross proceeds of $155,000. The aggregate net proceeds to the Company from the follow-on offering and concurrent private placement was $146,037 after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

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, transferability and conversion, as described below. As of March 31, 2021, no preferred shares were designated or issued.

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. The holders of Class A and Class B common shares, voting together as a single class, are entitled to elect the directors of the Company. Holders of Class A1 common shares and Class B1 common shares have no voting rights.

Dividends

The common shareholders are entitled to receive dividends, as may be declared by the Company's board of directors. Through March 31, 2021, no cash dividends have been declared or paid.

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 is convertible, at the holder's election 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 (subject to certain exceptions). Each Class B1 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 B1 common share is convertible into one Class A common share or one Class B common share at the holder's election (subject to certain exceptions). There are no conversion rights associated with the Class A common shares.

9. Share-Based Compensation

2018 Incentive Award Plan

In May 2018, the Company's board of directors and shareholders approved the 2018 Incentive Award Plan (the "2018 Plan"), which became effective on May 23, 2018. The 2018 Plan provides for the grant of incentive share options, nonqualified share options, share appreciation rights, restricted shares, dividend equivalents, restricted share units and other share- or cash- based awards. Upon the effectiveness of the 2018 Plan, the Company ceased granting awards under its 2015 Equity Incentive Plan (as amended, the "2015 Plan" together with the 2018 Plan, the "Plans").

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

A total of 4,466,500 Class A common shares were initially reserved for issuance under the 2018 Plan. The number of Class A common shares that may be issued under the 2018 Plan will automatically increase on each January 1, beginning in 2019 and continuing for each fiscal year until, and including, the fiscal year ending December 31, 2028, equal to the lesser of (1) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (2) a smaller number of Class A common shares determined by the Company's board of directors. In December 2020, the board of directors approved the automatic increase as of January 1, 2021 of 2,728,600 shares, equal to 4% of the as-converted Class A common shares outstanding on December 31, 2020. No more than 27,915,000 Class A common shares may be issued under the 2018 Plan upon the exercise of incentive options. The Class A common shares underlying any awards issued under the 2018 Plan or the 2015 Plan that on or after the effective date of the 2018 Plan expire, lapses unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised, or forfeited under the 2018 Plan or the 2015 Plan will be added back to the Class A common shares available for issuance under the 2018 Plan. As of March 31, 2021, 3,457,214 shares remained available for future grant.

2015 Equity Incentive Plan

Until May 23, 2018 (the effective date of the 2018 Plan), the 2015 Plan provided for the Company to grant incentive share options, nonqualified share options, share grants and other share-based awards to employees and non-employees to purchase the Company's Class A common shares. On the effective date of the 2018 Plan, the Company ceased granting awards under the 2015 Plan. At that time, the 4,691,213 Class A common shares subject to outstanding awards under the 2015 Plan remained reserved for issuance under the plan pursuant to such awards and the 92,170 Class A common shares that had been available for future grant under the 2015 Plan were no longer authorized and reserved for issuance or available for future grant under the 2015 Plan. The 2015 Plan continues to govern the terms and conditions of the outstanding awards granted under it. Class A common shares subject to awards granted under the 2015 Plan that expire, lapse unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised, or forfeited become available for issuance under the 2018 Plan. As of March 31, 2021, there were 2,758,352 Class A common shares subject to outstanding awards under the 2015 Plan and reserved for issuance there under pursuant to such awards.

2018 Employee Share Purchase Plan

In May 2018, the Company's board of directors and shareholders approved the 2018 Employee Share Purchase Plan (the "2018 ESPP"), which became effective on May 23, 2018. A total of 670,000 Class A common shares were initially reserved for issuance under the 2018 ESPP. The number of Class A common shares that may be issued under the 2018 ESPP will automatically increase on each January 1, beginning in 2019 and continuing for each fiscal year until, and including, the fiscal year ending December 31, 2028, equal to the lesser of (1) 1% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (2) a smaller number of Class A common shares determined by the Company's board of directors, provided that no more than 6,420,000 Class A common shares may be issued under the 2018 ESPP. In December 2020, the Company's board of directors approved an increase as of January 1, 2021 of 130,000 shares. As of March 31, 2021, 648,794 Class A common shares were available for future issuance under the 2018 ESPP.

Rilonacept Long-Term Incentive Plan

In December 2019, the compensation committee of the Company's board of directors approved the Company's Rilonacept Long-Term Incentive Plan ("RLTIP") under the 2018 Plan to incentivize eligible employees of the Company or any of its subsidiaries to achieve FDA approval for the commercial sale and marketing of rilonacept for recurrent pericarditis in the United States ("RLTIP Milestone"). The RLTIP provides for the potential to receive a cash award and two grants of restricted share units ("RSU") awards covering Class A common shares under the 2018 Plan. The target award value for each of the cash award and the two RSU awards will be equal to one-third of a participant's annual target bonus for the year of grant, as determined in accordance with the RLTIP. Depending on the date-range within

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

which the RLTIP Milestone is certified by the Compensation Committee as being achieved (such date the "Achievement Date"), the RLTIP provides for (1) an earnout percentage that can be achieved as to 100%, 50%, 25% or 0% and (2) an upside earnout percentage that can be achieved as to 50%, 25% or 0%. No awards will be earned or vest, and the second RSU award will not be granted, in the event the Achievement Date does not occur by a specified date. The cash award is eligible to be earned and vested upon the Achievement Date with respect to an amount determined based on the earnout percentage and the number of Class A common shares issuable under the first RSU award ("First RSU Award") upon the Achievement Date determined based on the earnout percentage and will vest on the first anniversary of the Achievement Date, subject to continued employment through such date. The second RSU award ("Second RSU Award") will be granted on the Achievement Date with respect to a number of shares determined based on both the earnout percentage and the upside earnout percentage, and will vest on the second anniversary of the Achievement Date, subject to continued employment through such date.

Options

Share option activity under the Plans is summarized as follows:

	Number of Shares	Weighted Average Fair Value
Outstanding as of December 31, 2020	9,958,858	$ 9.32
Granted	1,037,668	$ 15.14
Exercised	(115,012)	$ 6.62
Forfeited	(526,081)	$ 9.66
Outstanding as of March 31, 2021	10,355,433	$ 9.92
Share options exercisable as of March 31, 2021	4,355,393	$ 8.74
Share options unvested as of March 31, 2021	6,000,040	$ 11.43

Option Valuation

The assumptions that the Company used to determine the grant-date fair value of options granted to employees and directors under the Plans during the three months ended March 31, 2021 and 2020 were as follows, presented on a weighted-average basis:

	Three Months Ended March 31,	
	2021	2020
Risk-free interest rate	0.98 %	0.82 %
Expected term (in years)	6.25	6.25
Expected volatility	77.26 %	79.83 %
Expected dividend yield	— %	— %

Restricted Share Units

Restricted share units ("RSUs") represent the right to receive shares of the Company's Class A common shares upon vesting of the RSUs. The fair value of each RSU award is based on the closing price of the Company's Class A common shares on the date of grant.

In March 2021, the Company granted RSUs with service conditions ("Time-Based RSUs") to eligible employees. The Time-Based RSUs will vest 25% on each of the first, second, third and fourth anniversaries of the date of grant, subject to continued employment through such dates.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

In December 2019, the Company granted Time-Based RSUs that vested in one installment on December 31, 2020, subject to the recipient's continued employment through that date. As of December 31, 2020, 56,369 class A common shares were issued with the remaining shares 24,332 shares withheld for tax purposes.

During the year ended December 31, 2020 and in December 2019, the Company granted the First RSU Awards as part of the RLTIP to eligible employees. During the three months ended March 31, 2021, the Achievement Date was met and (1) the number of Class A common shares issuable under the First RSU Awards were determined in accordance with the RLTIP and will vest in one installment on the first anniversary of the Achievement Date, subject to continued employment through such date, and (2) the Second RSU Awards were granted to eligible employees on the Achievement Date with respect to a number of shares determined in accordance with the RLTIP and will vest on the second anniversary of the Achievement Date, subject to continued employment through such date.

During the three months ended March 31, 2021, the Company recognized compensation expense of $1,452 related to RSUs including those granted in connection to the RLTIP. During the three months ended March 31, 2020, the Company recognized $261 in compensation expense related to the Time-Based RSUs and the Company did not recognize any compensation expense related to the First RSU Award, as achievement of the RLTIP Milestone was determined to be not probable as of that date.

The following table summarizes RSU activity, including the RSUs outstanding under the RLTIP for the three months ended March 31, 2021:

	Number of Shares	Weighted Average Grant Date Fair Value
Unvested RSUs as of December 31, 2020	205,312	$ 13.41
Granted	395,903	$ 21.90
Vested	—	$ —
Forfeited	(17,630)	$ 12.92
Unvested RSUs as of March 31, 2021	583,585	$ 19.19

Share-Based Compensation

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

	Three Months Ended March 31,	
	2021	2020
Research and development expenses	$ 2,634	$ 1,769
Selling, general and administrative expenses	4,492	2,440
	$ 7,126	$ 4,209

10. License, Acquisition and Collaboration 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

vixarelimab and other antibodies covered by certain patent rights, including patents and other intellectual property rights, clinical data, know-how, and clinical drug supply. In addition, Biogen granted to the Company a non-exclusive, sublicensable, worldwide license to certain background patent rights related to the vixarelimab 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, including milestone payments of $4,000 and $10,000 paid during the year ended December 31, 2017 and the year ended December 31, 2019, respectively, each payment was 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 on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

The Company also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to the vixarelimab 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.

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 three months ended March 31, 2021 and 2020, the Company recorded research and development expense of $14 and $64, respectively, related to the annual maintenance fee in connection with the retained contracts.

Beth Israel Deaconess Medical Center License Agreement

In 2019, the Company exercised the call option under the stock purchase option agreement with Primatope Therapeutics, Inc. ("Primatope") and acquired all of the outstanding securities of Primatope (the "Primatope Acquisition"). As a result of the Primatope Acquisition, the Company acquired the rights to an exclusive license to certain intellectual property rights controlled by Beth Israel Deaconess Medical Center, Inc. ("BIDMC") to make, use, develop and commercialize KPL-404 (the "BIDMC Agreement"). Under the BIDMC 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. Under the BIDMC Agreement, the Company is obligated to pay an insignificant annual maintenance fee as well as clinical and regulatory milestone payments of up to an aggregate of $1,200 to BIDMC. The Company is also obligated to pay a low single-digit royalty on annual net sales of products licensed under the agreement.

During the three months ended March 31, 2021 and 2020, the Company did not record any research and development expense in connection with the BIDMC Agreement.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

Regeneron License Agreement

In September 2017, the Company entered into a license agreement (as amended, 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 was also obligated to make payments to Regeneron of up to an aggregate of $27,500 upon the achievement of specified regulatory milestones, which includes a $7,500 milestone achieved and paid in the fourth quarter of 2020 and a $20,000 milestone achieved and paid in the three months ended March 31, 2021. The $20,000 milestone was accounted for as an intangible asset and will be amortized over the life of the underlying asset. Related amortization expense will be recorded as cost of goods sold in the Company's consolidated statement of operations and comprehensive loss. During the three months ended March 31, 2021 and 2020, the Company did not incur any research and development expense directly related to milestones due under the Regeneron Agreement.

Under the Regeneron Agreement, the Company is solely responsible for all development and commercialization activities and costs in its territories. The Company is also responsible for costs related to the filing, prosecution and maintenance of certain licensed patent rights. In March 2021, the FDA approved the sBLA for ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older and the Company assumed the sales and distribution of ARCALYST for the approved indications in the United States, including CAPS and DIRA. The Company will evenly split profits on sales of ARCALYST with Regeneron, after deducting certain commercialization expenses subject to specified limits.

Pursuant to the Regeneron Agreement, in September 2017, the parties entered into a clinical supply agreement under which Regeneron agreed to manufacture product solely for the Company's use in development activities. Pursuant to the Regeneron Agreement, during the three months ended March 31, 2021, the parties entered into a commercial supply agreement under which Regeneron agreed to manufacture product for the Company's use, including for commercial sales. During the three months ended March 31, 2021 and 2020, the Company did not incur any research and development expense related to the purchase of drug materials under the clinical supply agreement. During the three months ended March 31, 2021, the Company recorded inventory of $2,189 related to the purchase of commercial product under the commercial supply agreement (see Note 4). As of March 31, 2021, the Company had non-cancelable purchase commitments under the commercial supply agreement (see Note 13).

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 with one year's written notice. The Company may also terminate the agreement with three months' written notice if the licensed product is determined to have certain safety concerns.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

MedImmune License Agreement

In December 2017, the Company entered into a license agreement (as amended from time to time, 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 to 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 $5,000 pass-through payment due upon the achievement of a specified clinical milestone event which was achieved in the fourth quarter of 2018. Also included is a milestone payment of $10,000 due upon the earlier to occur of a specified regulatory milestone and December 31, 2018, unless the MedImmune Agreement is earlier terminated by either party. During the year ended December 31, 2019, the Company made both the $5,000 and $10,000 previously accrued milestone payments in accordance with the MedImmune Agreement. 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. In July 2020, the Company entered into an amendment to the MedImmune Agreement to establish a new coronavirus field and defer the payment of certain development and regulatory milestones as applied to the new coronavirus field. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

The Company is solely responsible for all development, manufacturing, and commercial activities and costs of the licensed products, including clinical studies or other tests necessary to support the use of a licensed product. The Company is also responsible for costs related to the filing, prosecution and maintenance of the licensed patent rights.

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 three months ended March 31, 2021 and 2020, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

11. Net Loss per Share

The rights, including the liquidation and dividend rights, of the holders of Class A, Class B, Class A1 and Class B1 common shares are identical, except with respect to voting, transferability and conversion (see Note 8). As the liquidation and dividend rights are identical, losses are allocated on a proportionate basis, and the resulting net loss per

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

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:

	Three Months Ended March 31,	
	2021	2020
Numerator:		
Net loss attributable to common shareholders	$ (49,484)	$ (26,419)
Denominator:		
Weighted average common shares outstanding—basic and diluted	68,269,486	55,322,690
Net loss per share attributable to common shareholders— basic and diluted	$ (0.72)	$ (0.48)

The Company's unvested RSUs have been excluded from the computation of basic net loss per share attributable to common shareholders.

The Company's potentially dilutive securities, which include options and unvested RSUs, have been excluded from the computation of diluted net loss per share attributable to common shareholders as the effect would be to reduce the net loss per share attributable to common shareholders. Therefore, the weighted average number of common shares outstanding used to calculate both basic and diluted net loss per share attributable to common shareholders is the same. The Company excluded the following potential common shares, presented based on amounts outstanding at each period end, from the computation of diluted net loss per share attributable to common shareholders for the periods indicated because including them would have had an anti-dilutive effect:

	As of March 31,	
	2021	2020
Options to purchase common shares	10,355,433	8,990,872
Unvested RSUs	583,585	302,043
	10,939,018	9,292,915

12. Income Taxes

As an exempted company incorporated under the laws of 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 losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards will be available to the Company for those losses. The Company's wholly owned U.S. subsidiaries, Kiniksa US and Primatope, are subject to federal and state income taxes in the United States. The Company's wholly owned subsidiary Kiniksa UK, and its wholly owned subsidiaries, Kiniksa Germany, Kiniksa France, and Kiniksa Switzerland are subject to taxation in their respective countries. Certain of the Company's subsidiaries, primarily Kiniksa US, operate under cost plus arrangements.

The income tax rate for the three months ended March 31, 2021 varied from the Bermuda statutory rate of zero primarily due to income subject to United States taxation under the Kiniksa US cost-plus arrangements with the Company and Kiniksa UK, net of tax adjustments, and U.S. federal and state research tax credits. Income tax provision for the three months ended March 31, 2021 was $210 primarily due to the effective tax rate for Kiniksa US cost-plus arrangements partially offset by a discrete tax benefit primarily related to tax benefits from share-based compensation taxable events. Income tax benefit for the three months ended March 31, 2020 was $2,179 and includes a discrete tax benefit primarily related to tax benefits from share-based compensation taxable events.

KINIKSA PHARMACEUTICALS, LTD
NOTES TO CONSOLIDATED FINANCIAL STATEMENTS
(Amounts in thousands, except share and per share amounts)
(Unaudited)

Management examines all positive and negative evidence to estimate whether sufficient future taxable income will be generated to realize existing deferred tax assets. The Company previously determined it was more likely than not that a majority of our net deferred tax assets would not be realized and concluded that a valuation allowance was required, which eliminated the majority of our net deferred tax assets recorded in our balance sheet. In the future, if the Company believes that it is more likely than not that it will realize the benefit of these deferred tax assets, it will adjust the valuation allowance and recognize an income tax benefit. There are no material deferred tax assets in the other jurisdictions.

13. Commitments and Contingencies

License Agreements

The Company entered into license agreements with various parties under which it is obligated to make contingent and non-contingent payments (see Note 10).

Manufacturing Commitments

The Company entered into agreements with several contract manufacturing organizations to provide the Company with preclinical and clinical trial materials. The Company entered into a commercial supply agreement with Regeneron to provide both clinical supply and commercial product (see Note 10). As of March 31, 2021, the Company had committed to minimum payments under these agreements totaling $17,091, all which are due within one year.

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, officers and other key personnel 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, officers or other key personnel. 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 is not aware of any claims under indemnification arrangements that 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 March 31, 2021 or December 31, 2020.

Legal Proceedings

The Company is not a party to any litigation and does not have contingency reserves established for any litigation liabilities.

Item 2. Management's Discussion and Analysis of Financial Condition and Results of Operations.

The following discussion and analysis of our financial condition and results of operations should be read in conjunction with our unaudited consolidated financial statements and related notes included elsewhere in this Quarterly Report on Form 10-Q, or Quarterly Report, and our audited consolidated financial statements and related notes for the year ended December 31, 2020 included in our Annual Report on Form 10-K, or Annual Report. Some of the information contained in this discussion and analysis or set forth elsewhere in this Quarterly Report, including information with respect to our plans and strategy for our business, includes forward-looking statements within the meaning of Section 27A of the Securities Act of 1933, as amended, and Section 21E of the Securities Exchange Act of 1934, as amended, that involve risks and uncertainties. As a result of many factors, including those factors set forth in the risks identified in Part II-Item 1A "Risk Factors" section of this Quarterly Report and our other filings with the Securities and Exchange Commission, or SEC, our actual results could differ materially from the results, performance or achievements expressed in or implied by these forward-looking statements.

Overview

We are a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa's portfolio of assets, ARCALYST® (rilonacept), mavrilimumab, vixarelimab and KPL-404, are based on strong biologic rationale or validated mechanisms, target underserved conditions, and offer the potential for differentiation. These assets are designed to modulate immunological pathways across a spectrum of diseases.

ARCALYST is an interleukin-1α, and interleukin-1β cytokine trap. We received U.S. Food and Drug Administration, or FDA, approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older on March 18, 2021. Recurrent pericarditis is a painful inflammatory cardiovascular disease with an estimated U.S. prevalent population of approximately 40,000 patients seeking and receiving medical treatment. ARCALYST is commercially available through a distribution network comprised of several specialty pharmacies, which provide access across the United States. Upon the approval of the supplemental Biologics License Application, or sBLA, for ARCALYST in recurrent pericarditis, the scope of the license granted to Kiniksa expanded to include the previously approved indications for the treatment of Cryopyrin-Associated Periodic Syndromes, or CAPS, specifically familial cold autoinflammatory syndrome and muckle-wells syndrome, and for the treatment of Deficiency of IL-1 Receptor Antagonist, or DIRA, in adults and children 12 years of age and older in the United States and Japan. We are responsible for sales and distribution of ARCALYST in all approved indications in the United States. We will evenly split profits on sales of ARCALYST after deducting certain commercialization expenses, subject to specified limits, with Regeneron.

Mavrilimumab is an investigational monoclonal antibody inhibitor targeting granulocyte-macrophage colony stimulating factor receptor alpha, or GM-CSFRα. We are evaluating mavrilimumab for the potential treatment of giant cell arteritis, or GCA, a chronic inflammatory disease of the medium-to-large arteries with an estimated U.S. prevalence of approximately 75,000 to 150,000 patients. We conducted a global, randomized, double-blind, placebo-controlled Phase 2 proof-of-concept clinical trial for the study of mavrilimumab in GCA. The trial achieved both the primary and efficacy endpoint of time-to-first adjudicated GCA flare by Week 26 in all treated patients and the secondary efficacy endpoint of sustained remission at Week 26 in all treated patients with statistical significance. Additionally, while the trial was not powered for individual disease cohorts, there was a consistent trend of efficacy across the new onset and relapsing/refractory cohorts. In September 2020, the FDA granted Orphan Drug designation for mavrilimumab for the treatment of GCA. We are also evaluating mavrilimumab in severe coronavirus 2019 disease, or COVID-19, pneumonia and hyperinflammation. We are currently conducting a global, randomized, double-blind, placebo-controlled adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. In April 2021, we announced data from the Phase 2 portion of the trial in non-mechanically-ventilated patients with severe COVID-19 pneumonia and hyperinflammation receiving local standard of care. Non-mechanically ventilated patients treated with mavrilimumab demonstrated a reduction in mechanical ventilation and death at Day 29 pooled across dose levels. The trial achieved its primary efficacy endpoint of the proportion of patients alive and free of mechanical ventilation at Day 29. Mavrilimumab was well-tolerated and exhibited a favorable safety profile. The Phase 3 portion of the Phase 2/3 trial is ongoing and we are engaged with the FDA and other government agencies to identify pathways to potentially

accelerated availability of mavrilimumab as a therapeutic option for severe COVID-19 patients. We expect to provide next steps for the broader mavrilimumab development program, including GCA, in the second quarter of 2021.

Vixarelimab is an investigational monoclonal antibody that is designed to simultaneously inhibit the signaling of the cytokines interleukin 31, or IL-31, and oncostatin M, or OSM, by targeting their common receptor subunit, oncostatin M receptor beta, or OSMRβ. We are evaluating vixarelimab for the potential treatment of with prurigo nodularis, a chronic inflammatory skin condition with an estimated U.S. prevalence of approximately 300,000 patients. We are conducting a global, randomized, double-blind, placebo-controlled Phase 2b dose-ranging clinical trial of vixarelimab in prurigo nodularis. The Phase 2b clinical trial is expected to enroll approximately 180 patients experiencing severe pruritus. Patients will be randomized to receive vixarelimab or placebo subcutaneously once-monthly. The primary efficacy endpoint is the percent change from baseline in the weekly-average Worst-Itch Numeric Rating Scale, or WI-NRS, at Week 16. Key secondary efficacy endpoints include the proportion of patients achieving a greater-than-or-equal-to 4-point weekly-average WI-NRS reduction at Week 16 and the proportion of patients achieving a 0/1 score (clear/almost clear) on the prurigo nodularis-investigator's global assessment, or PN-IGA, at Week 16.

KPL-404 is an investigational monoclonal antibody designed to inhibit interaction of CD40 with CD154, or CD40 ligand, signaling, a well-known pathway that plays a critical role in regulating B cell proliferation and T cell activation as well as antibody production. We conducted a randomized, double-blind, placebo-controlled, single-ascending-dose Phase 1 clinical trial of KPL-404 in healthy volunteers to evaluate safety and pharmacokinetics well as receptor occupancy, or RO, and T-cell dependent antibody response, or T-cell Dependent Antibody Response, or TDAR, in these subjects. In May 2021, we reported final data from the trial. KPL-404 showed dose-dependent increases in concentration across cohorts. All dose escalations occurred as per protocol with no dose-limiting safety findings. KPL-404 was well-tolerated, and there were no serious adverse reactions. Subjects dosed with KPL-404 10 mg/kg IV showed full RO through at least Day 71 and complete suppression of TDAR after keyhole limpet hemocyanin, or KLH, challenge and re-challenge through at least Day 57. Subjects dosed with KPL-404 5 mg/kg SC showed full RO through Day 43 and suppression of TDAR after KLH challenge through at least Day 29. These data confirm and extend previously-reported 3 mg/kg IV cohort data, in which RO and suppression of TDAR after KLH challenge were demonstrated through Day 29. The 3 mg/kg IV dose level had previously demonstrated complete suppression of memory TDAR response to a re-challenge on Day 29. Anti-drug antibodies to KPL-404 were suppressed for at least 57 days at 10 mg/kg IV; the suppression of antibody responses to the drug itself is an independent indicator of target engagement and pharmacodynamic effect. We plan to initiate a Phase 2 proof-of-concept clinical trial in rheumatoid arthritis in the second half of 2021. The trial will evaluate safety and pharmacokinetics of KPL-404 with subcutaneous administration over 12 weeks. Rheumatoid arthritis was selected as a well-characterized autoimmune disease with decades of published clinical data across diverse mechanistic classes, allowing for objective evaluation in established endpoints. The pharmacokinetic lead-in of the planned trial supports characterization of chronic administration of KPL-404 in a patient population and provides optionality to evaluate the therapeutic potential of KPL-404 across a range of other autoimmune diseases with pathologies believed to be mediated by the CD40-CD154 pathway.

Our future success is dependent on our ability to successfully commercialize ARCALYST and to develop, obtain regulatory approval for and successfully commercialize one or more of our current or future product candidates. Upon approval from the FDA of the commercial marketing of ARCALYST in the United States for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older in March 2021, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States and will evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. However, as a company we have limited experience obtaining marketing approval for product candidates, commercializing a therapeutic, supporting sales, marketing, and distribution activities and maintaining applicable infrastructure for these activities either directly and/or through agreements with third parties; as a result we may not be successful in commercializing ARCALYST or any future approved product candidates, if any, thus potentially impairing the commercial potential of ARCALYST and our other product candidates to generate any revenue.

On May 18, 2020, we completed a follow-on offering of 2,760,000 Class A common shares, inclusive of the exercise of the underwriters' overallotment option at a public offering price of $18.25 and a concurrent private placement of 1,600,000 Class A1 common shares at an offering price of $18.25 per share for aggregate gross proceeds of $79.6 million. The aggregate net proceeds to us from the follow-on offering and concurrent private placement, inclusive

of the over-allotment option exercise, was $74.5 million after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

On July 24, 2020, we completed a follow-on offering of 5,952,381 Class A common shares, at a public offering price of $21.00 and a concurrent private placement of 1,428,572 Class A1 common shares at an offering price of $21.00 per share for aggregate gross proceeds of $155.0 million. The aggregate net proceeds to us from the follow-on offering and concurrent private placement was $146.0 million after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

We have incurred significant operating losses since inception. Our ability to generate product revenue sufficient to achieve profitability will depend heavily on the successful commercialization of ARCALYST and the development and eventual commercialization of one or more of our current or future product candidates, if approved. Our net losses were $49.5 million and $26.4 million for the three months ended March 31, 2021 and 2020, respectively. As of March 31, 2021, we had an accumulated deficit of $567.0 million. We expect to continue to incur significant operating losses for at least the next several years as we advance our product candidates through preclinical and clinical development and, ultimately, seek regulatory approval. In addition, we expect to continue to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution of ARCALYST. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates.

As a result, until such time as we can generate significant revenue from product sales of ARCALYST and one or more of our current or future approved product candidates, if ever, we expect to finance our operations through public or private securities offerings, debt financings or other sources, which may include licensing, collaborations or other strategic transactions or arrangements. We may be unable to raise additional funds or enter into such other transactions or arrangements when needed on favorable terms, or at all. If we fail to raise capital or enter into such transactions or arrangements as and when needed, we may have to significantly delay, scale back or discontinue the development and or of one or more of our current or future product candidates or delay our pursuit of potential in-licenses or acquisitions or scale back on commercialization activities for ARCALYST.

Because of the numerous risks and uncertainties associated with product development, including any impact from the COVID-19 pandemic, 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 successfully commercialize ARCALYST and generate product sales from one or more of our current or future product candidates, if approved, 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 March 31, 2021, we had cash, cash equivalents and short-term investments of $264.0 million. We believe that our existing cash, cash equivalents and short-term investments will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months from the date of issuance of the unaudited consolidated financial statements included in this Quarterly Report. 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.” Our future viability beyond that point is dependent on our ability to raise additional capital to finance our operations.

Components of Our Results of Operations

Revenue

As of March 31, 2021, we had not generated any revenue from any products. If our development efforts for our current or future product candidates are successful and result in additional regulatory approval, we may be able to generate revenue in the future from sales of ARCALYST and current or future product candidates, if approved.

Operating Expenses

Research and Development Expenses

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

- expenses incurred to conduct the necessary preclinical studies and clinical trials required to obtain regulatory approval;
- expenses incurred under agreements with contract research organizations, or CROs, that are primarily engaged in the oversight and conduct of our clinical trials and contract manufacturing organizations, or CMOs, that are primarily engaged to provide preclinical and clinical drug substance and product for our research and development programs for our product candidates;
- other costs related to acquiring and manufacturing preclinical and clinical trial materials, including manufacturing validation batches, as well as investigative sites and consultants that conduct our clinical trials, preclinical studies and other scientific development services;
- payments made in cash or equity securities under third-party licensing, acquisition and other similar 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 preclinical development, process development, manufacturing and clinical development activities. Our direct research and development expenses by program also include fees incurred under license, acquisition and other similar 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 activities as well as for managing our preclinical and clinical development, process development and manufacturing activities.

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

	Three Months Ended March 31,	
	2021	2020
	(in thousands)	
ARCALYST	$ 2,801	$ 4,037
Mavrilimumab	8,862	2,210
Vixarelimab	2,657	2,655
KPL-404	506	1,271
Unallocated research and development expenses	13,857	10,728
Total research and development expenses	$ 28,683	$ 20,901

Research and development activities are central to our business. Product candidates in later stages of clinical development generally have higher 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 be substantial over the next several years as we conduct our ongoing and planned clinical trials for mavrilimumab, vixarelimab and KPL-404, as well as conduct other preclinical and clinical development including regulatory filings for our current and future product candidates. As a result, our related personnel costs will increase, 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 other similar agreements to acquire the rights to our product candidates.

Upon approval from the FDA of the commercial marketing of ARCALYST in the United States for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older in March 2021, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States. However, as a company we have limited experience obtaining marketing approval our product candidates, commercializing a therapeutic, supporting a sales, marketing, distribution of therapeutic products, and maintaining applicable infrastructure for these activities either directly and/or through agreements with third parties; as a result we may not be successful in commercializing ARCALYST, or any future approved product candidates, if any, thus potentially impairing the commercial potential of ARCALYST and our other product candidates to generate any revenue. At this time, we cannot reasonably estimate or know the nature, timing and costs of the efforts that will be necessary to complete the clinical development of our current or future product candidates or when, if ever, material net cash inflows may commence from ARCALYST or any of our current or future product candidates, if approved. This uncertainty is due to the numerous risks and uncertainties associated with product development and commercialization, including the uncertainty of:

- the potential impact of the COVID-19 pandemic on our business, including our preclinical studies, clinical trials and operations;
- the scope, progress, outcome and costs of our research and preclinical development activities, clinical trials and other development activities;
- establishing an appropriate safety and efficacy profile with IND enabling and clinical studies;
- the successful enrollment and initiation, performance and completion of preclinical studies and 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;

- increasing clinical and commercial manufacturing capabilities or making arrangements with additional third-party manufacturers to successfully manufacture our product candidates;
- development and timely delivery of clinical-grade and commercial-grade drug formulations that can be used in our clinical trials and for commercial launch;
- supporting or establishing a sales, marketing and distribution infrastructure to commercialize ARCALYST or for our current or future products for which we may obtain marketing approval;
- successfully launching commercial sales of ARCALYST or of our current or future product candidates, if and when approved, whether alone or in collaboration with others;
- making milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreements;
- obtaining, maintaining, defending and enforcing patent claims and other intellectual property rights;
- significant and changing government regulation; and
- maintaining a continued acceptable safety profile of ARCALYST or our current or future approved product candidates following approval, if any.

Selling, General and Administrative Expenses

Selling, 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, information technology, medical affairs, commercial and support personnel functions. Selling, 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 continue to increase in the future as we continue to perform commercialization activities and increase our headcount to support our business objectives. We also anticipate that we will continue to incur significant costs associated with being a public company, including accounting, audit, legal, compliance and director and officer insurance costs as well as investor and public relations expenses, and that such costs will increase over time especially as we are now a large accelerated filer and are no longer permitted to rely on exemptions from certain requirements that are applicable to public companies that are not emerging growth companies or smaller reporting companies.

Interest Income

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

Income Taxes

As an exempted company incorporated under the laws of 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 are currently available to us for those losses, while our assets remain in Bermuda. Our wholly owned U.S. subsidiaries, Kiniksa Pharmaceuticals Corp., or Kiniksa US, and Primatope Therapeutics, Inc., or Primatope, are subject to federal and state income taxes in the United States. Our wholly owned subsidiary Kiniksa Pharmaceuticals (UK), Ltd., and its wholly owned subsidiaries, Kiniksa Pharmaceuticals (Germany) GmbH, Kiniksa Pharmaceuticals (France) SARL, and Kiniksa Pharmaceuticals GmbH are subject to taxation in their

respective countries. Our provision for income taxes relates mainly to U.S. taxable income, generated by our wholly owned subsidiary Kiniksa US.

Results of Operations

Comparison of the Three Months Ended March 31, 2021 and 2020

The following table summarizes our results of operations for the three months ended March 31, 2021 and 2020:

	Three Months Ended March 31,		
	2021	2020	Change
	(in thousands)		
Operating expenses:			
Research and development	$ 28,683	$ 20,901	$ 7,782
Selling, general and administrative	20,600	8,486	12,114
Total operating expenses	49,283	29,387	19,896
Loss from operations	(49,283)	(29,387)	(19,896)
Interest income	9	789	(780)
Loss before (provision) benefit for income taxes	(49,274)	(28,598)	(20,676)
(Provision) benefit for income taxes	(210)	2,179	(2,389)
Net loss	$ (49,484)	$ (26,419)	$ (23,065)

Research and Development Expenses

	Three Months Ended March 31,		
	2021	2020	Change
	(in thousands)		
Direct research and development expenses by program:			
ARCALYST	$ 2,801	$ 4,037	$ (1,236)
Mavrilimumab	8,862	2,210	6,652
Vixarelimab	2,657	2,655	2
KPL-404	506	1,271	(765)
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	9,734	7,361	2,373
Other	4,123	3,367	756
Total research and development expenses	$ 28,683	$ 20,901	$ 7,782

Research and development expenses were $28.7 million for the three months ended March 31, 2021, compared to $20.9 million for the three months ended March 31, 2020, an increase of $7.8 million. During the three months ended March 31, 2021, the increase in research and development expenses incurred related to the mavrilimumab Phase 2/3 clinical trial in COVID-19 pneumonia and hyperinflammation, offset by a decrease in the cost associated with our RHADSODY trial. The following includes additional information on our development programs.

The direct costs for our ARCALYST program were $2.8 million during the three months ended March 31, 2021, compared to $4.0 million during the three months ended March 31, 2020, a decrease of $1.2 million. The decrease in expenses incurred related primarily to the completion of RHAPSODY, our global, pivotal Phase 3 clinical trial in recurrent pericarditis, and transition to the long-term extension.

The direct costs for our mavrilimumab program were $8.9 million during the three months ended March 31, 2021, compared to $2.2 million during the three months ended March 31, 2020, or an increase of $6.7 million. The increase in expenses incurred related primarily to our Phase 2/3 clinical trial in COVID-19 pneumonia and hyperinflammation.

The direct costs for our vixarelimab program were $2.7 million during the three months ended March 31, 2021, compared to $2.7 million during the three months ended March 31, 2020. The costs were consistent during the three months ended March 31, 2021 and 2020. Expenses incurred during the three months ended March 31, 2021 was primarily related to the start-up of our Phase 2b clinical trial in prurigo nodularis while during the three months ended March 31, 2020 expenses were primarily related to the continued progress and expansion of our Phase 2a clinical trial in prurigo nodularis, as well as costs related to the conclusion of our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus.

The direct costs for our KPL-404 program were $0.5 million during the three months ended March 31, 2021, compared to $1.3 million during the three months ended March 31, 2020, a decrease of $0.8 million. The decrease in expenses incurred primarily related to limited clinical trial expenses for our Phase 1 trial of KPL-404 in healthy volunteers due to the completion of the patient portion of the clinical trial.

Unallocated research and development expenses were $13.9 million for the three months ended March 31, 2021 compared to $10.7 million for the three months ended March 31, 2020. The increase of $3.2 million in unallocated research and development expenses was due to an increase in personnel-related costs, related to share-based compensation. Personnel-related costs for the three months ended March 31, 2021 and 2020 included share-based compensation of $2.6 million and $1.8 million, respectively.

Selling, General and Administrative Expenses

Selling, general and administrative expenses were $20.6 million for the three months ended March 31, 2021 compared to $8.5 million for the three months ended March 31, 2020. The increase of $12.1 million was primarily due to an increase of $7.8 million in personnel-related costs and an increase of $3.2 million in costs associated with pre-commercialization activities for our ARCALYST program. Personnel-related costs for the three months ended March 31, 2021 and 2020 included share-based compensation of $4.5 million and $2.4 million, respectively.

Interest Income

Interest income was $9.0 thousand for the three months ended March 31, 2021 compared to $0.8 million for the three months ended March 31, 2020. The decrease was due primarily to lower interest rates on U.S. Treasury notes.

(Provision) Benefit for Income Taxes

For the three months ended March 31, 2021, we recorded a provision for income taxes of $0.2 million relating primarily to the tax impact from the current tax expense due to income from our cost plus arrangements in the United States, net of R&D credits utilized offset by tax benefit related to the exercise of share options. For the three months ended March 31, 2020, we recorded a benefit for income taxes of $2.2 million relating primarily to the tax impact from the exercise of share options.

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.

On May 18, 2020, we completed a follow-on offering of 2,760,000 Class A common shares, inclusive of the exercise of the underwriters' overallotment option at a public offering price of $18.25 and a concurrent private placement of 1,600,000 Class A1 common shares at an offering price of $18.25 per share for aggregate gross proceeds of $79.6 million. The aggregate net proceeds to us from the follow-on offering and concurrent private placement, inclusive of the over-allotment option exercise, was $74.5 million after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

On July 24, 2020, we completed a follow-on offering of 5,952,381 Class A common shares, at a public offering price of $21.00 and a concurrent private placement of 1,428,572 Class A1 common shares at an offering price of $21.00

per share for aggregate gross proceeds of $155.0 million. The estimated aggregate net proceeds to us from the follow-on offering and concurrent private placement was $146.0 million after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

As of March 31, 2021, we had cash, cash equivalents and short-term investments of $264.0 million.

Cash Flows

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

	Three Months Ended March 31,	
	2021	2020
	(in thousands)	
Net cash used in operating activities	$ (40,122)	$ (31,796)
Net cash provided by investing activities	44,834	88,790
Net cash provided by financing activities	1,106	2,414
Net increase in cash and cash equivalents and restricted cash	$ 5,818	$ 59,408

Operating Activities

During the three months ended March 31, 2021, operating activities used $40.1 million of cash, primarily resulting from our net loss of $49.4 million offset by net cash provided by our operating assets and liabilities of $0.9 million and non-cash charges of $8.4 million. Net cash provided by our operating assets and liabilities for the three months ended March 31, 2021 consisted primarily of a $1.0 million increase in accrued expenses and other liabilities primarily due to increases in related to our clinical trial costs and other general and administration accruals offset by our pre-commercialization activities of our ARCALYST program and the cash payment of the 2020 employee bonuses, offset by a $0.4 million decrease in operating lease liabilities due to monthly payments for our right-of-use assets.

During the three months ended March 31, 2020, operating activities used $31.8 million of cash, primarily resulting from our net loss of $26.4 million and net cash used in our operating assets and liabilities of $9.2 million, partially offset by non-cash charges of $3.8 million. Net cash used in our operating assets and liabilities for the three months ended March 31, 2020 consisted of a $1.2 million decrease in accounts payable and a $7.0 decrease in accrued expenses and other liabilities primarily due to the cash payment of the 2019 employee bonuses.

Investing Activities

During the three months ended March 31, 2021 investing activities provided $44.8 million of cash, consisting of $139.2 million from proceeds of maturities of short-term investments, partially offset by $74.4 million of purchases of short-term investments and $20.0 million related to the intangible asset acquired as a result of the milestone incurred under the Regeneron Agreement.

During the three months ended March 31, 2020 investing activities provided $88.8 million of cash, consisting of $115.2 million from proceeds of maturities of short-term investments, partially offset by $26.2 million of purchases of short-term investments and $0.2 million of purchases of property and equipment.

Financing Activities

During the three months ended March 31, 2021, net cash provided by financing activities was $1.1 million, consisting proceeds from the exercise of share options.

During the three months ended March 31, 2020, net cash provided by financing activities was $2.4 million, primarily consisting of proceeds from exercise of share options.

Funding Requirements

We expect to incur significant expenses in connection with our ongoing and planned activities as we commercialize ARCALYST and advance our current and future product candidates through preclinical and clinical development, seek regulatory approval and commercialize one or more of our current or future product candidates, if approved. We expect to continue to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution with respect to ARCALYST and, if we obtain marketing approval for any of our current or future product candidates, we expect to incur significant additional commercialization expenses related to such activities. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates. As a result, our related personnel costs will increase, 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 other similar agreements to acquire the rights to our product candidates. Additionally, we expect to continue to incur costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses. We expect to incur expenses as we:

- conduct our current and planned clinical trials for mavrilimumab, vixarelimab and KPL-404, as well as for any future product candidates, as applicable;
- increase clinical and commercial manufacturing capabilities or make arrangements with additional third party manufacturers to successfully manufacture our product candidates;
- develop and timely deliver clinical grade and commercial grade product formulations that can be used in our clinical trials and for commercial launch;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- maintain or establish a sales, marketing, medical affairs and distribution infrastructure to commercialize ARCALYST or any of our current or future product candidates for which we may obtain marketing approval and intend to commercialize on our own;
- launch commercial sales of ARCALYST and of any of our current or future product candidates, if and when approved, whether alone or in collaboration with others;
- make milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreements;
- hire additional clinical, quality and research and development personnel;
- expand our operational, financial and management systems and increase personnel globally 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
- in-license or acquire other product candidates and technologies or their related businesses, if we determine to do so.

We believe that our existing cash, cash equivalents and short-term investments 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 and technologies or their related businesses. We expect to continue to incur significant commercialization expenses related to product manufacturing, sales, marketing and distribution of ARCALYST. In addition, if we obtain

regulatory approval for any of our current or future product candidates, pursue additional indications for our products or any of our current or future product candidates, we expect to incur significant expenses related to product development and 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 products, 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:

- any impact of the COVID-19 pandemic on our business, including our preclinical studies and clinical trials, and operations;
- the scope, progress, results and costs of researching and developing our product candidates, and conducting preclinical 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 preclinical development efforts and our clinical trials and commercialization;
- the costs of future activities, including product sales, medical affairs, marketing, manufacturing, pricing and reimbursement, distribution and compliance, 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 and ongoing sales;
- the ability to receive additional non-dilutive funding;
- the revenue received from commercial sale of ARCALYST or any of our current or future products candidates, should they 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 licensing, collaboration or other strategic transactions and arrangements on favorable terms, if at all;
- the extent to which we acquire or in-license other product candidates, technologies and their related businesses; 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, or other sources, including, licensing, collaboration, marketing, distribution or other strategic transactions or arrangements with third parties. To the extent that we raise additional capital through the sale of equity or convertible debt securities, our shareholders' ownership interest may be materially diluted, and the terms of such securities could include liquidation or other preferences that adversely affect our shareholders' 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 licensing, collaboration, marketing, distribution or other strategic transactions or arrangements with third parties, we may have to relinquish valuable rights to our technologies, product candidates or future revenue streams, or otherwise agree to terms that may not be favorable to us. If we are unable to obtain funding, we could be forced to delay, reduce or eliminate some or all of our research and development programs for product candidates, product portfolio expansion or commercialization efforts, which could adversely affect our business prospects, or we may be unable to continue operations.

Contractual Obligations and Commitments

During the three months ended March 31, 2021, there were no material changes outside the ordinary course of our business to our contractual obligations and commitments set forth under the heading "Management's Discussion and Analysis of Financial Condition and Results of Operations—Contractual Obligations and Commitments" in the Annual Report. See Note 13 to our consolidated financial statements included in Item 1, "Consolidated Unaudited Financial Statements," of this Quarterly Report for a discussion of obligations and commitments.

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, revenue, 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.

During the three months ended March 31, 2021, there were no material changes to our critical accounting policies. Our critical accounting policies are described under the heading "Management's Discussion and Analysis of Financial Condition and Results of Operations—Critical Accounting Policies and Significant Judgments and Estimates" in the Annual Report and the notes to the consolidated financial statements included in Item 1, "Consolidated Unaudited Financial Statements," included in this Quarterly Report. We believe that of our critical accounting policies, the following accounting policies involve the most judgment and complexity:

- accrued research and development expenses; and
- share-based compensation.

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 SEC.

Item 3. Quantitative and Qualitative Disclosures About Market Risk.

Interest Rate Risk

We are exposed to market risk related to changes in interest rates. As of March 31, 2021, our cash, cash equivalents and short-term investments consisted of money market funds and U.S. Treasury notes. Our primary exposure to market risk is interest income sensitivity, which is affected by changes in the general level of U.S. interest rates. Further, the COVID-19 pandemic has adversely impacted the U.S. and global economy and financial markets, and any prolonged impact may have an impact on market interest rates. However, because of the short-term nature of the

instruments in our portfolio, an immediate 10% change in market interest rates would not have a material impact on the fair market value of our investment portfolio or on our financial position or results of operations.

Item 4. Controls and Procedures.

Limitations on Effectiveness of Controls and Procedures

In designing and evaluating our disclosure controls and procedures, management recognizes that any controls and procedures, no matter how well designed and operated, can provide only reasonable assurance of achieving the desired control objectives.

Evaluation of Disclosure Controls and Procedures

Our management, with the participation of our principal executive officer and principal financial officer, evaluated, as of the end of the period covered by this Quarterly Report, the effectiveness of our disclosure controls and procedures (as defined in Rules 13a-15(e) and 15d-15(e) under the Securities Exchange Act of 1934, as amended, or the Exchange Act). Based on that evaluation, our principal executive officer and principal financial officer concluded that our disclosure controls and procedures were effective at the reasonable assurance level as of March 31, 2021.

Changes in Internal Control over Financial Reporting

During the three months ended March 31, 2021, we completed the migration of our legacy accounting system to an SAP platform. In connection with this implementation, we updated the processes that constitute our internal control over financial reporting, as necessary, to accommodate related changes in our business processes. There were no other material changes to our internal control over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Securities Exchange Act of 1934) that occurred during the period covered by this Quarterly Report that have materially affected, or are reasonably likely to materially affect, our internal control over financial reporting.

PART II - OTHER INFORMATION

Item 1. Legal Proceedings.

We are not party to any material legal proceedings.

Item 1A. Risk Factors.

You should carefully consider the risks described below, as well as the other information in this Quarterly Report, including our unaudited consolidated financial statements and the related notes and "Management's Discussion and Analysis of Financial Condition and Results of Operations." 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. 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 biopharmaceutical company and have only started to generate 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.

Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk. Typically, it takes many years to develop one new product from the time it is discovered to when it is available for treating patients, and development may cease for a number of reasons. We have incurred operating losses in each year since our inception in 2015 and anticipate incurring losses for the foreseeable future. Our future success is dependent on our ability to develop, obtain regulatory approval for and successfully commercialize one or more of our product candidates. For example, while the U.S. Food and Drug Administration, or the FDA, approved ARCALYST® (rilonacept) for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021, we may not be able to successfully commercialize ARCALYST or any future product that may be approved.

We have incurred significant losses related to expenses for research and development and our ongoing operations. As of March 31, 2021, we had an accumulated deficit of $567.0 million. We expect to continue to incur losses for the foreseeable future, and anticipate these losses will increase substantially as a result of many factors, including:

- supporting our sales, marketing and distribution capabilities, infrastructure and organization to commercialize ARCALYST and any products candidates for which we may obtain marketing approval for indications in the United States;
- our research and preclinical and clinical development of our product candidates, including our global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in COVID-19, pneumonia and hyperinflammation, Phase 2b dose-ranging study of vixarelimab in prurigo nodularis, and planned Phase 2 clinical trial for KPL-404;
- manufacturing our products or product candidates for clinical or commercial use, and increasing our manufacturing capabilities or adding additional manufacturers or suppliers;
- seeking regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;

- initiating potential additional preclinical studies and clinical trials for our product candidates;
- making milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreement;
- seeking to identify, assess and study new or expanded indications for our products or product candidates, new or alternative dosing levels and frequency for our products or product candidates, or new or alternative administration of our products or product candidates, including method, mode or delivery device;
- seeking to identify, assess, acquire or develop additional product candidates;
- entering into licensing, acquisition, collaboration or other strategic transaction agreements;
- seeking to maintain, protect and expand our intellectual property portfolio;
- seeking to attract and retain skilled personnel;
- creating additional infrastructure to support our operations as a public company and large accelerated filer, our product development and commercialization efforts; and
- experiencing delays or encountering issues with any of the above, including but not limited to the impact of the COVID-19 pandemic and measures taken in response to the COVID-19 pandemic, failed trials, complex results, safety issues, regulatory challenges that require longer follow-up of existing trials, additional major trials, or additional supportive trials in order to pursue marketing approval.

See “Risk Factors — Risks related to product development — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact on our current or planned preclinical studies and clinical trials, which could be significant.”

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. 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 (deficit) 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, will force us to delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or other operations or commercialization efforts.

The development and commercialization of biopharmaceutical products is capital intensive. We are launching ARCALYST for the treatment of recurrent pericarditis and advancing our product candidates through research, preclinical and clinical development, including our global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation, our Phase 2b dose-ranging study of vixarelimab in prurigo nodularis, and our planned Phase 2 clinical trial with KPL-404.

We expect our expenses to increase in connection with our ongoing activities as we continue to support our sales, marketing and distribution capabilities, and continue the research and development of our product candidates, and expand our infrastructure and organization, and enter into agreements with third parties to conduct commercialization activities. Upon the FDA’s approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States and will evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. We expect to incur significant additional commercialization expenses with respect to our sales and marketing activities for ARCALYST as well as leading up to

or after any future marketing approval of any of our product candidates related to manufacturing, product sales, marketing and distribution. As our product candidates progress through development and towards potential commercialization, we will need to make milestone payments and, if successful, eventually make profit-split or royalty payments to the licensors and other third parties from whom we have acquired our product candidates. Furthermore, we expect to continue to incur costs associated with operating as a public company, including with respect to increased compliance and disclosure obligations as a result of becoming a large accelerated filer effective as of December 31, 2020 and no longer being an emerging growth company as of that time or a smaller reporting company as of January 1, 2021.

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 acceptable terms, if at all, we will be forced to delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or commercialization efforts. We also may not be able to expand our operations or otherwise capitalize on our business opportunities, or may be required to relinquish rights to our product candidates or products.

The commercialization and development process for our products and product candidates is highly uncertain, and we cannot estimate with certainty the actual amounts necessary to successfully market and sell products, or 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, including delays in or other adverse impacts to our commercialization of ARCALYST impacting our ability to generate projected revenue from ARCALYST or delays in the development or commercialization of any of our product candidates in the future, and we may need to seek additional funds sooner than expected, through public or private securities offerings, debt financings or other sources, including government funding or grants. Our future capital requirements will depend on, and could increase significantly as a result of, many factors, including:

- our ability to successfully commercialize ARCALYST or any of our product candidates if approved in the future, including the cost and timing of supporting our sales, marketing and distribution capabilities, infrastructure and organization expansion and entering into agreements with third parties to conduct one or more of these activities for commercialization of ARCALYST or for any of our product candidates, if approved or in anticipation of such approval in the future;

- the amount and timing of sales revenues from ARCALYST or any of our product candidates, if approved in the future, including the sales price and the availability of coverage and adequate third-party reimbursement;

- competitive and potentially competitive products and technologies and patients' and physicians' receptivity to ARCALYST or any of our product candidates if approved in the future and the technology underlying them in light of competitive products and technologies;

- the costs and timing of payments for producing ARCALYST or any of our product candidates to support clinical trials as well as the potential commercial launch of any of our product candidates if approved in the future, reserving manufacturing slots, or transferring manufacturing technology to third-party manufacturers;

- the results from, and the time and cost necessary for development of our product candidates, including for our global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation, our Phase 2b dose-ranging trial of vixarelimab in prurigo nodularis, and our planned Phase 2 clinical trial for KPL-404;

- the costs and timing of establishing and maintaining clinical trial sites for the development of our product candidates, both in the United States and in jurisdictions outside of the United States, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic;

- the number, size and type of preclinical activities and any additional clinical trials;

- the costs, timing and outcomes of seeking and potentially obtaining approvals from the FDA or comparable regulatory authorities outside of the United States, including the potential for the FDA or such comparable regulatory authorities to require that we conduct more studies than we currently plan 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 timing and amount of milestone and other payments we must make under our agreements with Regeneron, MedImmune, Limited, or MedImmune, Biogen MA Inc., or Biogen, 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;

- the cash requirements for seeking to identify, assess and study new or expanded indications for our products and product candidates, new or alternative dosing levels or frequency for our products or product candidates, or new or alternative administration of our products or product candidates, including method, mode or delivery device;

- the cash requirements of any future in-license, acquisition, development or discovery of additional product candidates, including in connection with any licensing, acquisition, collaboration or other strategic transaction 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 ARCALYST or our product candidates or any related activities;

- the costs associated with being a public company, including as a result of becoming a large accelerated filer as of December 31, 2020 and no longer being an emerging growth company at that time or a smaller reporting company as of January 1, 2021;

- our need and ability to hire and retain skilled personnel; and

- the receptivity of the capital markets to financings by biopharmaceutical companies generally and companies with a single commercial product and 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. The COVID-19 pandemic continues to adversely impact the global economy with the potential for the continued economic downturn to be severe and prolonged. A continued severe or prolonged economic downturn could result in a variety of risks to our business, including disruptions in the financial markets, that may make our ability to raise additional capital when needed, including through private or public securities offerings and debt financings more difficult to obtain, if at all, 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 when needed, we will be forced to curtail, delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or commercialization efforts of any of our products or product candidates for which we obtain approval. 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, restrict our operations or require us to relinquish rights to our technologies or product candidates or products.

Until such time as we can generate substantial product revenue, if ever, we expect to finance most of our cash needs through private or public securities offerings, debt financings, government funding or grants, or other sources, including licensing, collaboration or other strategic transactions or arrangements with third parties and to a lesser extent through projected revenue from ARCALYST or any of our product candidates, if approved in the future. 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. 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. Obtaining funds through licensing, collaboration or other strategic transactions or arrangements with third parties may require us 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 Class A common shares to decline.

Risks Related to Commercialization

We have limited experience as a company commercializing a therapeutic product and supporting sales, marketing, distribution and general infrastructure either directly and/or through agreements with third parties. As a company we have limited experience selling, marketing and distributing any therapeutic products. As a result we may not be successful in commercializing ARCALYST or any future approved product candidates, thus potentially impairing commercial potential for ARCALYST and our product candidates to generate any revenue.

The FDA approved ARCALYST for the treatment of recurrent pericarditis and reduction in risk of occurrence in adults and children 12 years of age and older in March 2021. Upon approval by the FDA, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States. While members of our management team have obtained marketing approvals for product candidates in the past while employed at other companies, we as a company have not previously obtained marketing approvals for any of our product candidates and have not previously sold, marketed or distributed any therapeutic products until now. To achieve commercial success for ARCALYST, or any future approved product candidate, we have established and expanded our internal capabilities, including but not limited to, sales, marketing, distribution, access and patient support services as well as making arrangements with third parties to perform certain services. We recruited and trained a specialty cardiology sales force of approximately 30 representatives who are charged with calling on high-volume accounts and specialists. We have hired other customer facing teams, including for our medical affairs, payor and patient support services teams to complement the efforts of the sales force, as well as developed an efficient digital marketing campaign. Our internal capabilities are augmented through contracts with third parties for distribution services, price reporting and aspects of our patient services programs. We undertook these efforts in order to commercialize ARCALYST in the United States. Each aspect of commercialization on its own can be complex, expensive and time consuming, and collectively the required effort for coordination is intensive. For example, recruiting and training a sales force and establishing marketing, payor and patient support service capabilities is expensive and time consuming and if not executed as planned could delay or reduce the effectiveness of the launch of ARCALYST and our product candidates, if approved.

Factors that may inhibit our efforts to timely and successfully commercialize ARCALYST or any of our current or future product candidates, if approved, include:

- our inability to recruit, train and retain adequate numbers of effective sales, marketing, access, and payor and patient support personnel;

- the inability of sales personnel to obtain access, especially within the restrictions of the ongoing COVID-19 pandemic, to physicians and accounts as well as for an adequate number of physicians or accounts to prescribe ARCALYST or any of our future products;

- the lack of complementary products to be supported by sales personnel, which may put us at a competitive disadvantage relative to companies with more extensive product lines;

- our inability to develop strong scientific-based relationships to drive disease awareness and education;

- our inability to establish the unmet medical need for the disease, and, with respect to ARCALYST or any of our future products, our inability to enable it to be viewed as the product of choice within the indication in which it is approved;

- our inability or delay in gaining reimbursement and broad patient access at a price that reflects the value of ARCALYST or any of our future products;

- our inability to equip customer-facing personnel with effective materials, including medical and sales literature to help them educate physicians and other healthcare providers regarding applicable diseases relevant to ARCALYST or any of our future products;

- our inability to effectively distribute products in a timely manner;

- our inability to provide physicians and patients adequate support and training to build comfort around reconstitution and self-administration process to initiate and continue to use ARCALYST or any of our future products;

- our inability to develop robust patient support programs to optimize the patient and customer experience with ARCALYST or any of our future products as well as with the Company;

- our inability to develop or obtain and sustain sufficient operational functions and infrastructure to support our commercial activities; and

- unforeseen costs and expenses associated with creating a sales, marketing, and access organization.

In addition, disruptions caused by the COVID-19 pandemic may increase the likelihood that we encounter such difficulties, delays or unforeseen costs. If we experience any of the factors that may inhibit our efforts to commercialize ARCALYST or any of our product candidates, if approved, we will not be successful in commercializing ARCALYST or any such future product candidates. Further, our business, results of operations, financial condition and prospects will be materially adversely affected.

The impact of the COVID-19 pandemic and measures taken in response to the COVID-19 pandemic has resulted in limitations on certain commercial activities, which, if prolonged, may impede the effective commercialization of ARCALYST or any of our product candidates, if approved, and result in lower than anticipated future revenue.

As part of our commercial strategy for ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence, we have implemented a focused and targeted launch effort, with a specialty cardiology salesforce of approximately 30 representatives charged with calling on high-volume accounts and high-volume specialists to help ensure that decision-makers view recurrent pericarditis as a serious, debilitating disease with a significant unmet need and view ARCALYST as the product of choice for the treatment of recurrent pericarditis and reduction in risk of recurrence.

The COVID-19 pandemic and measures taken in response to the COVID-19 pandemic, including business and travel restrictions and social-distancing to halt the spread of the pandemic, has had an impact on businesses, healthcare

systems, regulatory authorities and other organizations and conferences. These measures may result in limitations on certain aspects of our commercialization strategy, including our specialty cardiology salesforce not being able to access, or having limited access, to physician offices and other high-volume accounts in person, which if prolonged, may impede the effective commercialization of ARCALYST and result in lower than anticipated future revenue.

Our current products or future approved product candidates may not gain market acceptance by physicians, patients, or third-party payors (e.g., governments and private health insurers), in which case our ability to generate product revenues will be impaired.

Even with FDA or any other regulatory authority approval of the marketing of ARCALYST or any of our other product candidates in the future (whether developed on our own or with a collaborator), physicians, healthcare providers, patients, the medical community or third-party payors may not accept or use ARCALYST or any of our future product candidates, or may effectively block or limit their use in the case of third-party payors. If ARCALYST or any of our other product candidates, if approved, do not achieve an adequate level of acceptance, we may not generate the projected level of product revenue or profits from operations, if at all. The degree of market acceptance of ARCALYST in the approved recurrent pericarditis indication, or any of our future approved product candidates, if any, will depend on a variety of factors, including:

- the timing of market introduction;
- disease awareness, including understanding the severity and epidemiology of the disease;
- the number and clinical profile of competing products, whether regulatory approved or not;
- the potential and perceived advantages or disadvantages of our product candidates relative to alternative treatments;
- 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;
- convenience and ease of administration, including relative to alternative therapies;
- pricing (including patient out-of-pocket costs), budget impact, affordability and cost effectiveness, particularly in relation to alternative treatments;
- the effectiveness of our sales, marketing and distribution activities;
- availability of adequate coverage, reimbursement and payment from health maintenance organizations and other insurers, both public and private, and the timing thereof; and
- other potential advantages over alternative treatment methods.

If ARCALYST or any of our future approved products, if any, fail to gain market acceptance, our ability to generate revenue will be adversely affected. Even if ARCALYST or any future approved product candidates achieve market acceptance, the relevant market may prove not to be large enough to allow us to generate significant revenue.

The successful commercialization of our products and future approved product candidates, if any, will depend in part on the extent to which third-party payors, including governmental authorities and private health insurers, provide funding, establish favorable coverage and pricing policies and set adequate reimbursement levels. Failure to obtain or maintain coverage and adequate reimbursement for our products and future approved product candidates, if any, could limit our ability to market those products and decrease our ability to generate revenue.

Our ability to successfully commercialize ARCALYST in the approved recurrent pericarditis indication and other approved indications in the United States or any of our future approved product candidates, if any, particularly in orphan or rare disease indications, will depend in part on the availability of favorable coverage and the adequacy of reimbursement for ARCALYST or the product candidate and alternative treatments from third-party payors (e.g., governmental authorities, private health insurers and other organizations). We are seeking favorable coverage and reimbursement for ARCALYST for the treatment of recurrent pericarditis and reduction of risk of recurrence in adults and children 12 years of age and older from third-party payors. Obtaining coverage and adequate reimbursement is contingent on our ability to:

- obtain and present clinical data that supports payor value/benefit assessments;
- execute formal payor value/benefit assessment processes;
- obtain coverage that enables use in populations reflected in any product candidate's approved product label; and
- effectively negotiate favorable pricing and reimbursement terms.

While in some markets, there is a single payor, in other markets there are multiple payors that can have different ways of assessing prescription drugs and therapeutics. To successfully commercialize ARCALYST or any of our product candidates, if approved, we will be required to utilize the expertise, internally or through a third party, and resources that are sufficient to execute on the respective product candidate's coverage and reimbursement strategy. We cannot be certain we will be able to effectively execute our coverage and reimbursement strategy in the markets we pursue, which could limit the commercial potential of ARCALYST in the approved recurrent pericarditis indication or any of our product candidates, if approved. As a result, our ability to generate projected revenue from ARCALYST or any of our product candidates, if approved, could be negatively impacted.

Governmental authorities, private health insurers and other third-party payors have attempted to control costs by delaying the time to reimbursement, and by restricting the breadth of coverage and limiting the amount of reimbursement for particular products in terms of lower pricing and increasing the proportion of the cost for which the patient is responsible. There may be significant delays in obtaining reimbursement for newly approved products or product indications, coverage may be limited to a subset of the patient population for which the treatment is approved by the FDA or similar regulatory authorities outside the United States, and reimbursement rates may vary according to the use of the product and the clinical setting in which it is used. Coverage and reimbursement barriers by payors may materially impact the demand for, or the price of, , ARCALYST and any product candidate for which we obtain marketing approval, if any. If coverage and reimbursement are not available, or available only at limited levels, or if such coverage will require patient out-of-pocket costs that are unacceptably high, we may not be able to successfully commercialize , ARCALYST or any of product candidates for which we obtain marketing approval. Moreover, any coverage or reimbursement that may be obtained may be decreased or eliminated in the future.

Third-party payors continue to introduce new tactics to contain costs, including more rigorous value/benefit assessment processes and criteria. For example, it is possible that third-party payors will select low-cost clinical comparators that serve as benchmarks for determining relative value, including generics, biosimilars and lower costs brands with or without the same approved indication. The result of such a change would be a more challenging value/benefit assessment caused by a more challenging basis for comparison and the potential for a worse relative outcome. Third-party payors may determine that we have failed to generate sufficient evidence to demonstrate the relative benefits of ARCALYST or any of our product candidates, if approved, and refuse to provide coverage and reimbursement entirely, or many find the evidence not sufficiently compelling to support the desired pricing and

reimbursement. Similarly payors may implement coverage criteria that seeks to limit the use of ARCALYST or any of our product candidates, if approved, to situations where a patient must be proven to not adequately respond to the lower-cost comparator. The potential of third-party payors to introduce more rigorous value/benefit assessment processes and criteria could have a negative impact on our ability to commercialize ARCALYST or any of our product candidates for which we receive marketing approval successfully.

Third-party payors are also introducing more challenging price negotiation methodologies, including in re-visiting established coverage and reimbursement parameters when new competitors, including brands, generics and biosimilars enter the market. It is possible that a third-party payor may consider our product candidates as substitutable and only be willing to cover the cost of the alternative product. Even if we show improved efficacy, safety or improved convenience of administration with ARCALYST or any of our product candidates, if approved, pricing of competitive products may limit the amount we will be able to charge for ARCALYST or any of our product candidates, if approved. Third-party 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. In some cases, when new competitor generic and biosimilar products enter the market, there are mandatory price reductions for the innovator compound, in other cases, payors employ “therapeutic category” price referencing and seek to lower the reimbursement levels for all treatment in the respective therapeutic category. Additionally, new competitor brand drugs can trigger therapeutic category reviews in the interest of modifying coverage and or reimbursement levels. The potential of third-party payors to introduce more challenging price negotiation methodologies could have a negative impact on our ability to commercialize ARCALYST or any of our product candidates, if approved, successfully.

The incidence and prevalence for target patient populations of our products or product candidates have not been established with precision. If the market opportunities for our products and 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 not known with specificity, including with respect to COVID-19 pneumonia and hyperinflammation. 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 products and product candidates, are based largely on our extrapolation from available population studies and estimates. These estimates have been derived from a variety of sources, including scientific literature, surveys of clinics, patient foundations, pharmacy claims analyses, large national surveillance databases or market research, and may prove to be incorrect. Further, new trials and therapeutic options may lead to changes in the estimated incidence or prevalence of these diseases, or relevant subpopulations thereof, including the introduction of a vaccine for the prevention of COVID-19 with respect to COVID-19 pneumonia and hyperinflammation. As a result, the number of patients who may benefit from our products or product candidates may turn out to be lower than expected.

The total addressable market for any of our products and approved product candidates in the future, if any, will ultimately depend upon, among other things, the diagnostic criteria and applicable patient population included in the final label for the product or product candidate approved for sale for its indication, the efficacy, safety and tolerability demonstrated by the product candidate in our clinical trials, acceptance by the medical community and patients, pricing, access and reimbursement. The number of addressable patients in the United States and other major markets outside of the United States 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 significant market share.

Evolving health policy and associated legislative changes related to coverage and reimbursement aimed at lowering healthcare expenditure could impact the commercialization of our product candidates. Pharmaceutical pricing has been, and likely will continue to be, a central component of these efforts.

The regulations that govern regulatory approvals, pricing and reimbursement for new pharmaceutical products vary widely from country to country. In markets of some of the countries we may pursue outside of the United States for any of our product candidates, the products may be subject to extensive governmental price control or other price regulations. 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 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 negotiations that delay our commercial launch of the product candidate in that country, possibly for lengthy time periods, which may negatively impact the revenues we are able to generate from the sale of the product candidate in that country. Adverse pricing and reimbursement limitations may hinder our ability to recoup our investment in one or more product candidates, even if our product candidates obtain marketing approval.

Net prices for products may be reduced by mandatory discounts or legislated rebates that must be paid in order to participate in government healthcare programs or paid to other third-party payors. Mandatory discounts can be legislated at any time in any market. Similarly, some markets currently have pricing legislation that sets the price of a pharmaceutical product in their market by referencing the price of that product in other markets, known as international reference pricing. International reference pricing has the potential to impact price cut decisions in individual countries and the countries that reference the pricing of certain other individual countries. Expansion of mandatory discounts and international reference pricing, including into the United States, presents a material risk to our ability to achieve favorable pricing and adequate reimbursement.

Drug importation and cross-border trade, both sanctioned and unsanctioned, occurs when a pharmaceutical product from a market where the official price is set lower is shipped and made commercially available in a market where the official price is set higher. Any future relaxation of laws that presently restrict or limit drug importation or cross-border trade, including in the United States, could have a material negative impact on our ability to commercialize ARCALYST or any of our product candidates, if approved.

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, we may not be able to achieve or sustain favorable pricing for ARCALYST or any of our product candidates, if approved, and adequate reimbursement.

If, in the future, we are unable to maintain our sales, marketing and distribution capabilities, infrastructure and organization directly and/or through agreements with third parties to sell and market ARCALYST in recurrent pericarditis or any product candidates, if approved, the commercial potential for ARCALYST and our product candidates, if approved, to generate any revenue may be impaired.

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 drug 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.

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 approved product candidates ourselves. In addition, we may not be successful in entering into arrangements with third parties to market and sell our approved product candidates, if any, or may be unable to do so on terms that are favorable to us. Further, we will likely 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 approved product candidates effectively. However, developing a sales, marketing and access organization requires significant investment, is time consuming and if not completed as planned could delay the launch of our approved product candidates. Furthermore, we may not be able to adequately establish an effective sales, marketing, distribution and access organization in the EU or other key markets in

which we have obtained approval for the commercial marketing of our product candidates outside of the United States. If we are unable to maintain or reestablish sales, marketing and distribution capabilities successfully, either on our own or in collaboration with third parties, we will not be successful in commercializing our approved product candidates, if any, and approved product candidates ability to generate any revenue may be impaired. Furthermore, our business, results of operations, financial condition and prospects will be materially adversely affected.

Our future growth may depend, in part, on our ability to penetrate markets outside of the United States, 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 markets outside of the United States for which we may rely on collaborations with third parties.

Although we do not have immediate plans to pursue the regulatory approval and commercialization of ARCALYST for recurrent pericarditis or any other indication outside of the United States, we are evaluating the opportunities for the development and commercialization of our product candidates in certain markets outside of the United States. 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 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, manufacturing 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 markets outside of the United States, we would be subject to additional risks and uncertainties, including:

- our ability to obtain reimbursement for our product candidates in such markets;
- our inability to directly control commercial activities because we may rely on third parties;
- the burden of complying with complex and changing regulatory, tax, accounting and legal requirements of such countries;
- different medical practices and customs in such 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 such 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 laws of such country in the event of a contract dispute.

Sales of our product candidates outside of the United States 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, price negotiations with governmental authorities can take considerable time after the receipt of marketing approval for a drug. To obtain adequate reimbursement or favorable pricing approval in some countries, we may be required to conduct a clinical trial that compares 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.

We are subject to ongoing obligations, regulatory requirements and continued regulatory review, which may result in significant additional expense. Additionally, our products and future approved product candidates, if any, 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.

We are subject to ongoing regulatory requirements for manufacturing, labeling, packaging, distribution, storage, advertising, promotion, sampling, record-keeping, AE reporting, conduct of post-marketing trials and submission of safety, efficacy and other post-market information for our products, including both federal and state requirements in the United States. We are also subject to additional ongoing obligations and continued regulatory review, which may result in significant additional expense. Furthermore, if we seek and receive approval from comparable regulatory authorities outside of the United States for products or any of our product candidates, if approved, in the future, we will be subject the requirements of comparable regulatory authorities outside of the United States.

Manufacturers and their facilities are required to comply with extensive requirements of the FDA and comparable regulatory authorities outside of the United States, 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 our CMOs 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 regulatory authorities outside of the United States. Any new legislation addressing drug safety issues could result in delays in product development or commercialization, or increased costs to assure compliance.

While our current clinical and medical affairs activities are subject to certain ongoing regulatory requirements concerning appropriate exchange of medical and scientific information, they must also comply with additional 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 our product, such as AEs 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 AEs 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, co-marketers or other third-parties operating on our behalf 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 the 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 previous U.S. presidential administration took 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 whether or how these Executive Orders will be implemented by the current U.S. presidential administration, if at all, and the extent to which they may 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. Further, the policies and priorities of the new administration are unknown and could materially impact the regulations governing our product candidates. 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 be subject to enforcement actions 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 and other healthcare provider payment and price transparency, 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.

Upon the FDA's approval of ARCALYST for the treatment of recurrent pericarditis and the reduction in risk of recurrence in adults and children 12 years of age and older in March 2021, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States and will evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. As we assumed the sales and distribution responsibilities of ARCALYST for the approved indications in the United States and began marketing ARCALYST in the approved recurrent pericarditis indication, we became subject to additional healthcare statutory and regulatory requirements and enforcement by the United States federal and state governments and the governments of other countries or jurisdictions in which we conduct our business.

Healthcare professionals, physicians and third-party payors play a primary role in the recommendation and prescription of ARCALYST and any product candidates for which we obtain marketing approval. Our 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 ARCALYST and 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. 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 False Claims Act even when they do not submit claims directly to government payors if they are deemed to "cause" the submission of false or fraudulent claims. Moreover, the government may assert that a claim including items and services resulting from a violation of the federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the False Claims Act;

- 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 service. 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 6002 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 certain financial interactions with physicians (defined to include doctors, dentists, optometrists, podiatrists and chiropractors), additional categories of healthcare practitioners beginning in 2022, and teaching hospitals, as well as the ownership and investment interests of physicians and their immediate family members; 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 professionals or marketing expenditures and pricing information.

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, prescribers or other potential purchasers of our products or 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 prescribers to be in violation of applicable laws or the appearance of a conflict of interest. For example, 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. The FDA or other regulatory authorities may conclude that a financial relationship between us and a principal investigator or a clinical trial site has created a conflict of interest or otherwise affected interpretation of a study. The FDA or other regulatory authorities 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, which could result in a delay in approval, or rejection, of our marketing applications by the FDA or other regulatory authorities and may ultimately lead to the denial of marketing approval of our product candidates. Furthermore, investigators for our clinical trials may become debarred by FDA or other regulatory authorities, which may impact the integrity of our studies and the utility of the clinical trial itself may be jeopardized. 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 healthcare professionals are also governed by strict laws, regulations, industry self-regulation codes of conduct and healthcare professionals' codes of professional conduct in Europe and individual European member states. The provision of any inducements to healthcare professionals to prescribe, recommend, endorse, order, purchase, supply, use or administer a drug product is prohibited. A number of European member states have established additional rules requiring pharmaceutical companies to publicly disclose their interactions with physicians and other healthcare professionals and to obtain approval from employers, professional organizations or competent authorities before entering into agreements with healthcare professionals.

Ensuring that our business arrangements with third parties comply with applicable healthcare laws and regulations may 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 activities 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 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.

Risks Related to Product Development

We depend heavily on the success of one or more of our products and product candidates, 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 successfully commercialize one of our product candidates, or experience significant delays in doing so, our business will be significantly harmed.

We are not permitted to market or promote any of our product candidates before we receive regulatory approval from the FDA or comparable regulatory authorities outside of the United States. Our product candidates are in various stages of clinical development. Our assumptions about why our product candidates are worthy of future development and potential approval in the indications for which we are studying them, or any other indications, are based in part on indirect data collected by other companies and in part from data collected from our preclinical and clinical trials. 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 Mavrilimumab was studied in Phase 2 clinical trials conducted by MedImmune outside of the United States for the treatment of rheumatoid arthritis, or RA, we studied mavrilimumab in a global Phase 2 clinical trial for the treatment of GCA, for which we announced that the trial met both the primary and secondary efficacy endpoints with statistical significance, and are conducting a global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation, for which we released top-line data from the first cohort of the Phase 2 portion of the trial, and are enrolling and dosing patients in a Phase 3 portion of the trial. We have been studying vixarelimab in prurigo nodularis, for which we released top-line data from our Phase 2a clinical trial, and are enrolling and dosing patients in a Phase 2b dose-ranging study of vixarelimab in prurigo nodularis. In addition, we are planning to initiate a Phase 2 clinical trial of KPL-404. Our future preclinical product candidates would need to progress through toxicology studies and other requirements to enable an Investigational New Drug application, or IND, prior to clinical development. We cannot be certain that any of our product candidates will be successful in these clinical trials or will receive regulatory approval even after completing a successful pivotal clinical trial. We may also determine that the potential product and commercial profile of any of our product candidates may not ultimately be commercially successful, or even if they have the potential to be commercially successful, we may not have sufficient resources, which in either case could lead us to discontinue development of one or more of our product candidates or we may otherwise determine to not support further development of any of our product candidates at any time for any reason. If we do not receive regulatory approvals for more than one of our product candidates, we may not be able to continue our operations.

While we received FDA approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older, each of our product candidates require additional preclinical or clinical development and all of our product candidates will require regulatory approval in one or more jurisdictions, manufacturing capacity and expertise, successful manufacture of clinical supply, and, if ultimately approved, will require an organization to support commercialization and product launch, and significant marketing efforts before we will be able to generate any revenue from product sales of such product candidates, if approved. The success of our product candidates or potential future product candidates depends upon several factors, including the following:

- submission to and acceptance by the FDA of INDs and of clinical trial applications to governmental authorities outside of the United States for our product candidates to commence planned clinical trials or future clinical trials;

- successful completion of preclinical 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;

- successful site activation for, enrollment in, and completion of clinical trials, the design and implementation of which are agreed to by the applicable regulatory authorities, and the ability of our 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 and maintenance of any such approvals;

- as applicable, pediatric study plans acceptable to the FDA and comparable regulatory authorities outside of the United States, and follow through of any pediatric study commitments, including development of pediatric formulations where indicated;

- establishment and maintenance of arrangements with third-party manufacturers, as applicable, for continued clinical supply and commercial manufacturing;

- successful development of our manufacturing processes and transfer to 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;

- successful manufacture of sufficient supplies of our product candidates within approved specifications for purity and efficacy from our facility and from our CMOs or other sole-source manufacturers in order to meet clinical or commercial demand, as applicable;

- establishment and maintenance of patent and trade secret protection or regulatory exclusivity for our product candidates;

- timely and successful commercial launch of our product candidates;

- acceptance of our products, if and when approved, by patients, patient-advocates, 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 trial commitments or REMS; and

- maintenance of a continued acceptable safety profile of our product candidates following approval.

If we do not accomplish one or more of these factors in a timely manner or at all, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic, we could experience significant delays in, or an inability to, timely or successfully commercialize our product candidates, which would materially harm our business. If

we do not receive regulatory approvals for one or more additional product candidates, we may not be able to continue our operations. Even though we received FDA approval for ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older, and assumed the sales and distribution of ARCALYST for the previously approved indications in the United States, we will evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits, and the relevant markets for these indications may prove not to be large enough to allow us to generate significant revenue from these product sales. Moreover, even if we successfully obtain regulatory approvals to manufacture and market one or more 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, among other things. If the markets for patient subsets that we are targeting are smaller than we estimate, we may not generate projected revenue levels from sales of such product candidates, if approved.

Clinical drug development is a lengthy and expensive process with uncertain timelines and 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 successfully 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 our product candidates in humans. Clinical testing is expensive, time consuming and uncertain as to outcome.

Not all of our clinical trials have been conducted as initially planned or completed on our initial projected schedule, and accordingly, we cannot guarantee that any of our current or potential future clinical trials will be conducted as initially planned or completed on our initial projected schedule, if at all, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic.

Commencing a clinical trial is subject to acceptance by the FDA of an IND or IND amendments, acceptance by European regulatory authorities of a Clinical Trial Application, or CTA, or acceptance by other applicable regulatory authorities, and finalizing the trial design based on discussions with the FDA, European regulatory authorities or other applicable regulatory authorities. We have and may in the future receive feedback or guidance from regulatory authorities on our clinical trial design and protocols and, even after we incorporate such feedback or guidance from these regulatory authorities, such regulatory authorities may impose other requirements for our clinical trials, could disagree that we have satisfied their requirements to commence our clinical trials, disagree with our interpretation of data from the relevant preclinical studies, clinical trials or chemistry, manufacturing and controls, or CMC, data, or disagree or change their position on the acceptability of our trial designs, including the proposed dosing level or schedule, treatment duration, our definitions of the patient populations or the clinical endpoints selected, which may require us to complete additional preclinical studies, clinical trials, CMC development, other studies or impose stricter approval conditions than we currently expect.

For example, the FDA has provided feedback 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 option. In addition, we anticipate that other potential indications for mavrilimumab would need to be in serious or life-threatening diseases where the burden of the disease is sufficient to justify the risk-benefit of mavrilimumab to pursue clinical development in such indications or potentially in indications requiring limited doses where the theoretical risk of pulmonary alveolar proteinosis, or PAP, is low. Further, based on FDA feedback we received in connection with its review and approval of an IND for our global Phase 2 clinical trial of mavrilimumab in GCA, we anticipate that to help inform the risk-benefit profile for the use of mavrilimumab in GCA, we will need to eventually demonstrate safety and effectiveness of mavrilimumab beyond 26 weeks, as well as evaluate its pharmacokinetic profile and the effectiveness of mavrilimumab at different doses in GCA.

Commencing our planned clinical trials is also subject to approval by an IRB at each clinical trial site before a trial may be initiated, which approval could be delayed, rejected or suspended. Further 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 may impose a suspension or termination of our clinical trials even after approval and initiation of trial sites 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, unforeseen safety issues or adverse side effects that arise in the trial, failure to demonstrate a benefit from using a drug, any of which could result in the imposition of a clinical hold, as well as changes in governmental regulations or administrative actions or lack of adequate funding to continue the clinical trial.

Successful completion of our clinical trials is a prerequisite to submitting a Biologics License Application, or BLA, sBLA or New Drug Applications, or NDA, to the FDA and a marketing authorization application, or MAA, to the European Medicines Agency, or EMA, or other applicable regulatory authorities in other countries for each product candidate and, consequently, to obtaining approval and initiating commercial marketing of our current and any future product candidates. A failure of one or more of our current or future clinical trials can occur at any stage of testing, and our clinical trials may not be successful. We have and may continue to experience delays in our ongoing clinical trials, and we do not know whether planned clinical trials will begin on time, be allowed by regulatory authorities, need to be redesigned, or if we can activate sites or enroll patients on time, or if they will be completed on schedule, if at all. Events that have and may continue to delay or prevent commencement or successful completion of clinical development of our product candidates as planned and on schedule, if at all, include but are not limited to:

- inability to generate sufficient preclinical, toxicology or other in vivo or in vitro data to support the initiation of human clinical trials;
- delays or failure in reaching a consensus with regulatory agencies on trial design or implementation;
- delays or failure in establishing the appropriate dosage levels or frequency of dosing or treatment period in clinical trials;
- delays or failure in reaching 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 IRB approval at each clinical trial site;
- delays or failure in obtaining regulatory approval to commence a trial, or imposition of a clinical hold by regulatory authorities, after review of an IND or IND amendment, or equivalent application or amendment, or an inspection of our clinical trial operations or study sites;
- challenges in recruiting and enrolling suitable patients or a sufficient number thereof to participate in our clinical trials;
- amendments to clinical trial protocols impacting study criteria, endpoints or design, including amendments that either we initiate or are requested by regulatory authorities;
- difficulty collaborating with patient groups and investigators;
- failure by our CROs, medical institutions, other third parties we contract with in connection with our clinical trials, 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 comparable regulatory guidelines in other countries;

- patients not completing participation in a clinical trial or returning for post-treatment follow-up, in either case including as a result of trial demands on participants as a result of the COVID-19 pandemic and measures taken in response to the pandemic or otherwise, among other things;
- clinical trial sites or patients withdrawing from a clinical trial, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic or otherwise, among other things;
- participating patients experiencing serious adverse events or undesirable side effects or being exposed to unacceptable health risks;
- participating patients failing to experience confirmed pre-specified events during the clinical trial within an expected time-frame, if at all;
- safety issues, including occurrence of adverse events associated with the product candidate that are viewed to outweigh its potential benefits;
- difficulty in identifying the patient 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, inconclusive or uncompetitive results, which may result in us deciding, or regulators requiring us, to conduct additional clinical trials or modify or cease development programs for our product candidates;
- suspensions or terminations of our clinical trials by us or the IRBs of the institutions in which our clinical trials are being conducted, the Data Safety Monitoring Board for such trials or the FDA or comparable regulatory authorities;
- failure of manufacturers, or us, to produce phase-appropriate supplies of our product candidates for use in our clinical trials in accordance with current good manufacturing practices, or cGMP, requirements and regulations or applicable comparable regulatory guidelines in other countries;
- 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; and
- disruptions to our business operations, including our manufacturing operations, and the business operations of our third-party manufacturers, CROs upon whom we rely to conduct our clinical trials, or other third parties with whom we conduct business or otherwise engage, as well as disruptions in travel into and within the countries in which we conduct our clinical trials, our manufacturers produce our product candidates or we otherwise conduct business or engage with other third parties, now or in the future as a result of the impact of the COVID-19 pandemic.

Delays in the commencement or completion of our planned and ongoing clinical trials of our product candidates have occurred and may continue to occur. Consequences of delays have increased and may in the future increase our costs of developing our product candidates, slow down the development and approval of our product candidates, delay or jeopardize our ability to commence product sales and generate revenue, if any, from our product candidates and harm their commercial prospects. Furthermore, disruptions caused by the COVID-19 pandemic have increased and may continue to increase the likelihood that we encounter such difficulties or delays in commencing or completing our planned and ongoing clinical trials or other development. In addition, many of the factors that cause, or lead to, a

difficulties and delays in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates or us deciding to modify or cease development of our product candidates.

Clinical trial delays could also shorten any periods during which our products have patent protection or shorten any periods during which we have the exclusive right to commercialize our product candidates and may allow our competitors to bring products to market before we do, which could impair our ability to obtain orphan exclusivity for our products that potentially qualify for this designation and to successfully commercialize our product candidates, and may harm our business and results of operations. Any inability to successfully complete preclinical 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.

Furthermore, clinical trials must be conducted in accordance with the laws, rules and regulations, guidelines and other requirements of the FDA, European Union, or EU, and other applicable regulatory authorities outside of those jurisdictions and are subject to oversight by these regulatory authorities and IRBs at the medical institutions where the clinical trials are conducted. Further, conducting global clinical trials, as we do for certain of our product candidates, may require that we coordinate among the legal requirements and guidelines of regulatory authorities across a number of jurisdictions, including the United States, EU and countries outside of those jurisdictions, which could require that we amend clinical trial protocols or determine not to conduct a trial in one or more jurisdictions or to run separate trials in various jurisdictions due to the inability, cost or delay in harmonizing divergent requests from such regulatory authorities, all of which could increase costs. In addition, clinical trials that are conducted in countries outside the United States and the EU may subject us to risks associated with the engagement of non-United States and non-EU CROs who are unknown to the FDA or the EMA, and may have different standards of diagnosis, screening and medical care, as well as risks associated with further delays and expenses as a result of increased shipment costs (including as a result of local quality release or in-country testing of a product candidate supply produced in a different jurisdiction for our clinical trials) and political and economic risks relevant to such countries outside the United States and the EU.

The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact on our current or planned preclinical studies and clinical trials, which could be significant.

The COVID-19 pandemic, and measures taken in response to the pandemic, have had and could continue to have an impact on our current or planned preclinical studies and clinical trials. If the COVID-19 pandemic and measures undertaken in response to the pandemic are prolonged, or the easing of any of such measures has adverse consequences, we may experience significant disruptions that could materially impact our preclinical studies and clinical trials, including by:

- impeding, delaying, limiting or preventing the production, delivery or release of our product candidates to our clinical trial sites or patients, including due to interruptions in the supply of raw materials or global shipping that may affect the transport of our product candidates or clinical trial materials, or the reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19;

- impeding, delaying, limiting or preventing the production, delivery or release of the supply of our product candidates, including due to disruptions at manufacturing facilities that produce our product candidates, staffing shortages, reprioritizations, production slowdowns or stoppages or interruptions in global shipping;

- impeding, delaying, limiting or preventing clinical trial investigators, other critical staff, or patients from traveling to our clinical trial sites or visiting nurses traveling to patients;

- impeding, delaying, limiting or preventing key clinical trial activities, including clinical trial site monitoring, patient dosing, study procedures (such as biopsies, which may be deemed non-essential), collection of clinical data and samples as well as cleaning and verification of clinical data, which could affect the integrity of clinical trial data;

- diverting healthcare resources away from the conduct of clinical trials or reprioritizing the focus of such resources on clinical trials for product candidates with the potential for treatment or prevention of COVID-19 related conditions;

- timing of COVID 19 and other vaccinations received by potential patients for our clinical trials may impede, delay, limit or prevent such potential patients from enrolling in our clinical trials;

- impeding, delaying, limiting or preventing clinical trial site initiation, including difficulties in recruiting clinical site investigators and clinical site staff, and enrollment or retention of patients in our clinical trials;

- increasing the risk that participants enrolled in our clinical trials will acquire COVID-19 while the clinical trial is ongoing, which could impact the results of the clinical trial, including by increasing the number of observed adverse events;

- interrupting or delaying preclinical studies due to restricted or limited operations at our research and development laboratory facility;

- causing interruptions or delays at the FDA, or other regulatory authorities, which could result in delays in review and approval of our submissions and applications, including INDs, clinical trial protocols and BLAs for our product candidates;

- resulting in the refusal of the FDA to accept data from clinical trials in affected geographies;

- prompting changes in local regulations as part of a response to the COVID-19 pandemic which may require us to change the ways in which our clinical trials are conducted, which may result in unexpected costs, or cause us to pause or discontinue one or more of our current or planned clinical trials altogether;

- delaying necessary interactions with local regulators, ethics committees and other important agencies and contractors due to limitations in employee resources or forced furlough of government employees; and

- limiting employee resources that would otherwise be focused on the conduct of our clinical trials, including because of sickness of employees or their families or the desire or requirement of employees to avoid contact with large groups of people.

Any one of the foregoing could significantly impede, delay, limit or prevent the clinical development of our product candidates and ultimately lead to the delay or denial of regulatory approval of our product candidates. While we continuously look to identify business-critical activities and to develop contingencies and mitigation strategies for those activities to potentially minimize the impact of the COVID-19 pandemic on our business and operations, there can be no assurance that we will be able to identify all such activities or that any identified contingencies and mitigation strategies will be effective. If the clinical development of our product candidates is significantly impeded, delayed, limited or is prevented, it could ultimately lead to the delay or denial of regulatory approval of our product candidates which would materially adversely affect our business and operations, including our ability to generate revenue.

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, our particular enrollment criteria or competing clinical studies in the same patient population, including patients with COVID-19, or due to the impact of the COVID-19 pandemic. Difficulty in enrolling patients could delay or prevent completion of our 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 a sufficient number of patients to participate in testing our product candidates, including for the treatment of conditions associated with COVID-19, particularly given that many of the conditions for which we are evaluating our current product candidates or

may evaluate them in the future are in small disease populations. In addition, the eligibility criteria 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 evaluate based on the primary and secondary endpoints of the study. Further, our product candidates modulate the immune system and carry risks associated with immunosuppression, including the risk of serious infections, potential interference with vaccines, and other potential serious health risks.

Our clinical trials have and may continue to 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. For example, the large number of competitive studies for the treatment of conditions associated with COVID-19 and the evolving standard of care with overall improvement in the quality of care for patients diagnosed with COVID-19 initially had an impact on the number of eligible patients for our global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. 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 would reduce the number of patients who are available for our clinical trials at such clinical trial site.

In addition, disruptions to our business operations and the business operations of our CROs or other third parties with whom we conduct business, as well as disruptions in travel into and within the countries in which we conduct our clinical trials, now or in the future, as a result of the impact of the COVID-19 pandemic, may delay or prevent patient enrollment.

Accordingly, when we encounter these or other difficulties in enrollment we have and may continue to experience delays or we may be prevented from completing our clinical trials. Patient enrollment depends on many factors, including:

- the size and nature of the patient population;
- the severity of the disease being studied;
- patient referral practices of physicians;
- patient eligibility criteria for the clinical trial and evolving standards of care;
- the proximity of patients to clinical sites;
- the complexity of the design and nature of the clinical protocol and trial;
- the availability and nature of competing clinical trials;
- the availability of standard of care or new drugs approved for the indication the clinical trial is investigating;
- failure to obtain and maintain or timely amend patient consents;
- our ability to recruit clinical trial investigators with applicable competencies and experience;
- the risk that patients enrolled in clinical trials will withdraw from the trials before completion of their treatment or follow-up period (in either case including as a result of trial demands on participants among other things);
- clinicians’ and patients’ perceptions as to the safety and potential advantages of the product candidate being studied in relation to other available therapies; and

- the occurrence of adverse events, or AEs, or undesirable side effects attributable to our product candidates.

The process of finding and enrolling patients may prove costly, especially since we are looking to identify a subset of the patients eligible for our studies from a relatively small patient population for many of the diseases we are studying, including for the treatment of conditions associated with COVID-19. If patients are unable or unwilling to participate in our clinical trials for any reason, or we experience difficulties in patient enrollment for any other reasons, such as due to the COVID-19 pandemic, our costs may significantly increase and the timeline for recruiting patients, conducting trials and obtaining regulatory approval of our product candidates may be significantly delayed or prevented, the commercial prospects of our product candidates may be harmed, and our ability to commence product sales and generate product revenue from any of these product candidates could be delayed or prevented. Any of these occurrences may harm our business, financial condition, and prospects significantly.

Our products and 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 products and 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 more restrictive labels or the delay or denial of regulatory approvals by the FDA or other comparable regulatory authorities outside of the United States.

Our products and product candidates modulate the immune system and carry risks associated with immunosuppression, including the risk of serious infections and other potential serious health risks.

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 granulocyte macrophage colony stimulating factor, or GM-CSF, function. The disease can range in severity from a sub-clinical reduction in diffusion capacity to significant dyspnea during mild exertion. In preclinical 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 RA. Preclinical 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.

However, if the results of our clinical trials reveal an unacceptable severity and prevalence of these or other side effects, the FDA or applicable regulatory authority outside of the United States may suspend or terminate our clinical trials, or not authorize us to initiate further trials. In addition, if other anti-GM-CSF molecules in development by third parties show these or similar side effects, it could have an impact on the entire class of anti-GM-CSF molecules in development and the applicable regulatory agency may suspend or terminate our clinical trials, or not authorize us to initiate further trials. The FDA or comparable regulatory authorities outside of the United States could order us to cease further development of, or deny or withdraw any approval of, any of our products or product candidates for any or all targeted indications.

For example, we anticipate that other potential indications for mavrilimumab would need to be in serious or life-threatening diseases where the burden of the disease is sufficient to justify the risk-benefit of mavrilimumab to be studied in such indications or potentially in indications requiring limited doses where the theoretical risk of PAP is low. In addition, based on FDA feedback we received in connection with its review and authorization of an IND for our global Phase 2 clinical trial of mavrilimumab in GCA, we anticipate that to help inform the risk-benefit profile for the use of mavrilimumab in GCA, we will need to eventually demonstrate the safety and effectiveness of mavrilimumab beyond 26 weeks as well as evaluate its pharmacokinetic profile and the effectiveness of mavrilimumab at different doses in GCA.

In addition, the development of any of our product candidates in other potential indications could increase the possibility of identification of adverse safety results that impact our development of such product candidates. For example, the development of mavrilimumab in other potential indications, such as COVID-19 pneumonia and

hyperinflammation, could increase the possibility of identification of adverse safety results that impact our development of mavrilimumab for GCA or any other indication.

Further, clinical trials by their nature utilize a sample of the potential patient population. Certain rare and severe side effects associated with our products or product candidates may only be uncovered with a significantly larger number of patients exposed to the product candidates. For our products or any of our product candidates that receive marketing approval, and we or others later identify undesirable side effects caused by the 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 it 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 registry or 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 or other elements to assure safe use;
- we may be required to change the way the product is administered, conduct additional clinical trials or change the labeling of the product;
- 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 or product candidate, if approved, and could significantly harm our business, results of operations and prospects.

If we cannot replicate positive results from earlier preclinical studies and clinical trials conducted by us or third parties, including 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 current or future product candidates.

Positive results from our preclinical studies and any positive results we may obtain from our earlier clinical trials of our product candidates, or from the clinical trials conducted by third parties, including investigator-initiated studies or the companies from whom we in-licensed or acquired or may in the future in-license or acquire our product candidates, may not be predictive of the results from any required later preclinical studies and clinical trials. Similarly, the positive results from the preclinical studies and earlier clinical trials or investigator initiated studies of our product candidates may not be replicated in our subsequent preclinical studies and clinical trial or investigator initiated study results. The mechanisms of action of our product candidates may not prove to be safe or effective to treat the diseases we are studying. Further, the safety and efficacy of our product candidates have not been determined for the indications in which we are developing them, and we cannot provide any assurance that their development will be successful. For example, although mavrilimumab has been studied in Phase 2 clinical trials for RA, GCA and COVID-19, its safety and efficacy have not been established in the indications we are pursuing, and, if even any such indications are further pursued by us, we may fail to receive regulatory approval for those indications.

Many companies in the pharmaceutical and biotechnology industries have suffered significant setbacks in later-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, preclinical findings made while clinical trials were underway or safety or efficacy observations made in preclinical studies and clinical trials, including AEs previously unreported in earlier studies and trials and favorable safety and efficacy observed in earlier studies and trials not replicated in later studies or trials. Further, such setbacks may be caused by manufacturing or formulation changes to product candidates or changes in manufacturers or manufacturing processes to produce products as compared to the process or manufacturing methods used in prior preclinical studies and clinical trials. Moreover, preclinical and clinical data are often susceptible to varying interpretations and analyses and many companies that believed their product candidates performed satisfactorily in preclinical studies and clinical trials nonetheless failed to obtain FDA or EMA approval. Furthermore, the approval policies or regulations of the FDA or the applicable regulatory authorities outside of the United States may significantly change in a manner rendering our clinical data insufficient for approval, which may lead to the FDA or such other regulatory authorities delaying, limiting or denying approval of our product candidates.

Interim, preliminary, and "top-line" data from our clinical trials that we announce or publish from time to time may change as more patient data become available following the interim data; preliminary data are subject to audit and verification procedures, and deeper analysis of the data beyond the topline data may provide more color and context to the data, all of which could result in material or other changes in the final data.

From time to time, we may disclose interim data from our preclinical studies and clinical trials, which are based on an interim analysis of then-available data from ongoing studies or trials. Interim data from our preclinical studies and clinical trials that we may complete are subject to the risk that one or more of the clinical observations may materially change as patient enrollment continues and more patient data become available from the particular study or trial. As a result, interim data should be viewed with caution until final data are available. Adverse differences between interim data and final data could significantly harm the development of our product candidate and our business prospects with respect thereto.

Further, from time to time we may announce or publish preliminary data from our pre-clinical studies or clinical trials, which are based on a preliminary analysis of final data. Preliminary data from our preclinical studies and clinical trials are subject to change following a more comprehensive review of the data from the particular preclinical study or trial. We also make assumptions, estimations, calculations and conclusions as part of our preliminary analyses of the data, and we may not have received, or had the opportunity to evaluate fully and carefully, all of the data. As a result, preliminary data remain subject to audit and verification procedures that may result in the final data being different from the preliminary data we previously announced or published.

From time to time, we may also announce or publish topline data from our preclinical studies and clinical trials, which are a subset of the total data intended to provide the important results from the study or trial. As a result, deeper analysis of the data beyond the topline data may provide more color and context to the results. Any adverse color and context provided by the broader data to the topline data could significantly harm the development of our product candidate and our business prospects with respect thereto.

Further, third parties, including regulatory agencies, may not accept or agree with our assumptions, estimates, calculations, conclusions or analyses or may interpret or weigh the importance of data differently, which could impact the value of the particular program, the approvability or commercialization of the particular product candidate and our business prospects. In addition, the information we announce or publish regarding a particular preclinical study or clinical trial may represent only a portion of extensive information generated from that study or trial, and our shareholders or other third parties may not agree with what we determine is material, important or otherwise appropriate information to include in our disclosure.

If the interim, preliminary, or topline data that we report differ materially from final results, or if third parties, including regulatory authorities, disagree with the conclusions reached, our ability to obtain approval for, and commercialize, our product candidates may be harmed, which could harm our business prospects, operating results or financial condition. Further, announcement of preliminary, interim or top-line data by us or differences between that data and the final data could result in volatility in the price of our Class A common shares.

Risks Related to Marketing Approval and Regulatory Matters

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 current or future product candidates or we fail or otherwise cease to advance their development, we will be delayed in commercializing or will not be able to commercialize, our current or future product candidates and our ability to generate additional revenue will be materially impaired.

Our current or future product candidates and the activities associated with their development and commercialization, including their trial design, testing, manufacture, safety, efficacy, record-keeping, labeling, storage, approval, advertising, promotion, pricing, 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 current or future product candidates, we must obtain marketing approval. We may not be able to receive approval or clearance to market any of our current or future 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 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 preclinical 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 authorities. Our current or future 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, including 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. In addition to the United States, we may seek regulatory approval to commercialize our product candidates in other jurisdictions. 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.

The process of obtaining regulatory approvals, both in the United States and in other countries, is time consuming, expensive, may take many years, 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 preclinical studies or clinical or other trials for our current or future product candidates. Our current and future product candidates could be delayed in receiving, or fail to receive, regulatory approval or we may fail or cease to advance their development for many reasons, including the following:

- the FDA or comparable regulatory authorities in other jurisdictions 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 regulatory authorities in other jurisdictions that a product candidate is safe and effective for its proposed indication;
- the FDA or comparable regulatory authorities in other jurisdictions could require us to collect additional data or conduct additional clinical trials;
- we may be unable to demonstrate to the satisfaction of the FDA or comparable regulatory authorities in other jurisdictions that we or our CMOs can manufacture clinical trial material that is deemed to be comparable to the material used in previous clinical trials of our product candidates;

- the results of clinical trials may produce negative, inconclusive or uncompetitive results, which may result in us deciding, or regulators requiring us, to conduct additional clinical trials or to modify or cease development programs for our product candidates;
- the results of clinical trials may not meet the primary or secondary endpoints of the applicable trial or the level of statistical significance required by the FDA or comparable regulatory authorities in other jurisdictions;
- we may be unable to demonstrate that a product candidate's clinical and other benefits outweigh its safety risks;
- the FDA or comparable regulatory authorities in other jurisdictions may disagree with our interpretation of data from preclinical studies or clinical trials;
- the FDA or comparable regulatory authorities in other jurisdictions may disagree that we have provided sufficient safety data or adequately demonstrated clinical benefit in a patient population or subpopulation studied in the clinical trial;
- the data collected from clinical trials of our product candidates may not be sufficient to support the submission of a BLA, sBLA or other submission or to obtain regulatory approval in the United States or elsewhere;
- the FDA or comparable regulatory authorities in other jurisdictions could require us to conduct additional clinical trials to compare our product candidates to other therapies for the treatment of the same indication;
- 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 data quality and regulatory requirements or meet their contractual obligations to us in a timely manner, or at all;
- the FDA or comparable regulatory authorities in other jurisdictions 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 FDA or comparable regulatory authorities in other jurisdictions may not believe that their on-site inspections and data audits have sufficiently demonstrated the quality and integrity of the clinical trial conduct and of data submitted to the FDA or comparable regulatory authorities in other jurisdictions in support of our new product approvals and marketing applications;
- 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, toxicities or other unexpected characteristics, causing us or our investigators, regulators or IRBs to reject, suspend or terminate the clinical trials; and
- the approval policies or regulations of the FDA or comparable regulatory authorities in other jurisdictions may significantly change in a manner rendering our clinical data, biologic manufacturing process and other supporting information insufficient for approval.

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

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

Our products, current product candidates and any of our future product candidates regulated as biologics in the United States may face biosimilar 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 approved under a BLA 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, another company may still market a competing version of the reference product for the same therapeutic indication if the FDA approves a full BLA for the competing product containing the sponsor's own preclinical 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.

For example, although ARCALYST was approved as a biological product under a BLA for the treatment of CAPS in February 2008, and we believe it qualified for the 12-year period of exclusivity against any biosimilars, such 12-year period of exclusivity has lapsed. The FDA approved ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021. However, the 12-year exclusivity period does not attach to the approval of an sBLA, potentially creating the opportunity for biosimilar competition, subject to any orphan drug exclusivity under the U.S. Orphan Drug Act (See "Risk Factors — Risks Related to Marketing Approval and Regulatory Matters — We may seek orphan drug designation for our product candidates in the United States, as well as for any of our product candidates in the EU, 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."). If we obtain FDA approval for any of our other biological product candidates, we expect any such product candidates to qualify for the 12-year period of exclusivity under the BPCIA. However, there is a risk that this exclusivity could be shortened due to Congressional action or otherwise, or that the FDA will not consider any such approved product candidates to be reference products for competing products, potentially creating the opportunity for generic competition sooner than anticipated.

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 authorization of our current or future product candidates in a major pharmaceutical market such as the United States, or the EU, we may not seek or obtain approval or commercialize our current products or 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. Regulatory requirements can vary widely from country to country, and 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, additional administrative review periods, and additional preclinical studies or clinical trials, which would be costly and time consuming and could delay or prevent the introduction of our current or future product candidates, or ARCALYST, 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 our product candidates in the United States, as well as for any of our product candidates in the EU, 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.

We received orphan drug designations in the United States for ARCALYST for the treatment of pericarditis, which includes recurrent pericarditis, and for mavrilimumab for the treatment of GCA, and we may seek orphan drug designation for certain of our other product candidates in the United States as well as for any of our product candidates in the EU. We may be unsuccessful in obtaining such designation for any of our other product candidates or unable to maintain the associated benefits for ARCALYST now that it is approved by the FDA for recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older or for mavrilimumab or any of our other current or future product candidates that are granted orphan drug designation, if any. 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 EU, 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 EU, 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 EU 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 drug 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 EU would be sufficient to justify the necessary investment in developing the drug. In the EU, 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 us, 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 EU. The EU 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 the 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 received orphan drug designations in the United States for ARCALYST for the treatment of pericarditis and for mavrilimumab for the treatment of GCA, and we may seek orphan drug designation for our other current or future product candidates, we may never receive such designation for such other product candidates. Even though we received such designation for ARCALYST and mavrilimumab and may receive such designation for any of our other current or future product candidates, there is no guarantee that we will enjoy the benefits of such designations.

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

We received Breakthrough Therapy designation for ARCALYST for the treatment of recurrent pericarditis, and we may seek Breakthrough Therapy or Fast Track designation for some of our other 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. In addition, if a product candidate is intended for the treatment of a serious or life-threatening condition and clinical or preclinical 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 Fast Track and Breakthrough Therapy designations, and even if we believe a particular product candidate is eligible for this designation, we cannot be certain that the FDA would decide to grant it. Even if we obtain such designations 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 or Breakthrough Therapy designations if it believes that such designations are no longer supported. Although products receiving Fast Track and Breakthrough Therapy designation are generally eligible for the FDA's priority review procedures, receiving such designations does not guarantee that the BLA for such products will receive priority review.

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 candidate no longer meets the conditions for qualification.

We have limited experience obtaining marketing approvals, and we may be unable to successfully do so for any of our current or future product candidates. Failure to successfully complete another pivotal clinical trial or obtain marketing approval in a timely manner for any of our current or future product candidates could have a material adverse impact on our business and financial performance.

Conducting pivotal clinical trials and preparing, and obtaining marketing approval for, a product candidate is a complicated process. As a company, we have only limited experience in obtaining marketing approval for our product candidates. As a result, in the future, obtaining marketing approval for our any of current or future product candidates

may require more time and expense than we anticipate. Failure to successfully complete, or delays in, any of our eventual other pivotal trials or related regulatory submissions would prevent us from, or delay us in, obtaining regulatory approval for, or clearance of, our current or future product candidates. It is possible that the FDA or other regulatory authorities may refuse to accept for substantive review any regulatory submissions that we submit for our product candidates or may conclude after review of our applications for any of our current or future product candidates that the submissions are insufficient to obtain marketing approval or clearance of any of our current or future product candidates. If the FDA or other regulatory authorities do not accept our applications for our current or future product candidates, if any, or the FDA delays or does not issue marketing authorizations for any of our current or future product candidates, the FDA or other regulatory authorities may require that we conduct additional clinical, preclinical or manufacturing validation trials and submit that data before they will reconsider our applications. Depending on the extent of these or any other required trials, approval or receipt of any marketing authorization 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 or other regulatory authorities to approve or grant marketing authorizations. Any delay in obtaining, or an inability to obtain, marketing approvals would delay or prevent us from commercializing any of our current or future product candidates, generating additional revenue and achieving and sustaining profitability. If any of these outcomes occur, we may be forced to modify or cease our development efforts for one or more of our product candidates, which could significantly harm our business.

Disruptions at the FDA and other government agencies caused by funding shortages or global health concerns could hinder their ability to hire, retain or deploy key leadership and other personnel, or otherwise prevent new or modified products from being developed, approved or commercialized in a timely manner or at all, which could negatively impact our business.

The ability of the FDA to review and approve new products can be affected by a variety of factors, including government budget and funding levels, statutory, regulatory, and policy changes, the FDA's ability to hire and retain key personnel and accept the payment of user fees, and other events that may otherwise affect the FDA's ability to perform routine functions. Average review times at the FDA have fluctuated in recent years as a result. In addition, government funding of other government agencies that fund research and development activities is subject to the political process, which is inherently fluid and unpredictable. Disruptions at the FDA and other agencies may also slow the time necessary for new drugs and biologics or modifications to approved drugs or biologics to be reviewed and/or approved by necessary government agencies, which would adversely affect our business. For example, over the last several years, including for 35 days beginning on December 22, 2018, the U.S. government has shut down several times and certain regulatory agencies, such as the FDA, have had to furlough critical FDA employees and stop critical activities.

Separately, in response to the COVID-19 pandemic, on March 10, 2020 the FDA announced its intention to postpone most inspections of foreign manufacturing facilities, and on March 18, 2020, the FDA temporarily postponed routine surveillance inspections of domestic manufacturing facilities. Subsequently, on July 10, 2020 the FDA announced its intention to resume certain on-site inspections of domestic manufacturing facilities subject to a risk-based prioritization system. The FDA intends to use this risk-based assessment system to identify the categories of regulatory activity that can occur within a given geographic area, ranging from mission-critical inspections to resumption of all regulatory activities. Additionally, on April 15, 2021, the FDA issued a guidance document in which the FDA described its plans to conduct voluntary remote interactive evaluations of certain drug manufacturing facilities and clinical research sites. According to the guidance, the FDA intends to request such remote interactive evaluations in situations where an in-person inspection would not be prioritized, deemed mission-critical, or where direct inspection is otherwise limited by travel restrictions, but where the FDA determines that remote evaluation would be appropriate. Regulatory authorities outside the United States may adopt similar restrictions or other policy measures in response to the COVID-19 pandemic. If a prolonged government shutdown occurs, or if global health concerns continue to prevent the FDA or other regulatory authorities from conducting their regular inspections, reviews, or other regulatory activities, it could significantly impact the ability of the FDA or other regulatory authorities to timely review and process our regulatory submissions, which could have a material adverse effect on our business.

Risks Related to Manufacturing and Our Reliance on Third Parties

We contract with third parties for manufacturing our commercial supply of ARCALYST and clinical supply for our product candidates and for certain research and other preclinical development and expect that we will continue to do so in the future. This reliance on third parties increases the risk that we may not have sufficient quantities of ARCALYST or 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 late-stage or commercial manufacturing facilities. Although we have built a development and manufacturing facility to produce drug substance to support certain research, preclinical and other clinical development for our product candidates, we rely, and expect to continue to rely, on third parties for the manufacture of our late-stage product candidates and certain of our early-stage product candidates for the majority of our clinical development efforts, as well as for the commercial manufacture of ARCALYST or any of our current or future product candidates, if approved, as well as label and packaging activities. We rely on these third parties to produce ARCALYST and 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 ARCALYST and our product candidates or that ARCALYST and our product candidates are not produced at an acceptable cost or quality, which could delay, prevent or impair our development or commercialization efforts.

For example, Regeneron is the sole manufacturer of ARCALYST and we have a contract with Regeneron to produce ARCALYST on an exclusive basis for a period of time. However, Regeneron is not obligated to accept our forecasts or our purchase orders that are not in line with accepted forecasts and Regeneron may not have sufficient manufacturing capacity to meet our commercial demand of ARCALYST. Regeneron, in turn, relies upon a CMOs or other third-parties to conduct fill/finish operations for ARCALYST. Under certain circumstances, we or Regeneron could initiate a technology transfer to either us or another CMO to manufacture ARCALYST. Finding new CMOs or third-party suppliers to produce ARCALYST would add additional cost and require significant time and focus of our management team. The CMO would need to produce ARCALYST at a different manufacturing site and potentially using a different process or at a different scale. We cannot provide any assurance that the technology transfer from Regeneron to us or another CMO will be successful in producing ARCALYST in sufficient quantities or of acceptable quality, if at all, or that we or another CMO will produce a comparable product to the satisfaction of the FDA or other comparable regulatory authorities, which could delay, prevent or impair the further development, if any, or commercialization of ARCALYST. In addition, there is typically a transition period when a new CMO commences work. Any significant delay or interruption in the supply of ARCALYST by Regeneron or otherwise could considerably impact our ability to meet commercial or clinical demand for the product and our ability to generate revenue from ARCALYST could be materially impaired.

We also have CMOs manufacture vixarelimab drug substance and drug product and entered into an agreement with a CMO to produce mavrilimumab beyond our current inventory. While we have built a manufacturing facility to support early development for our product candidates, we and our CMOs may not be able to produce sufficient quantities of our product candidates or produce them at an acceptable quality, including as a result of the COVID-19 pandemic, which could delay, prevent or impair our development or commercialization efforts and increase costs.

As a result of the COVID-19 pandemic, existing and any new third-party CMOs or suppliers may be unable to produce or supply ARCALYST or our current or future product candidates or to obtain the raw materials needed to produce or supply of ARCALYST or our product candidates or may experience delays, restrictions or limitations in the production, delivery or release of the supply of them or the raw materials needed to produce them, including due to disruptions at the respective facilities that produce ARCALYST or our product candidates or obtain the raw materials needed to produce them, staffing shortages production slowdowns, stoppages or reprioritizations, including as a result of reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, or interruptions in global shipping. In addition, there is typically a transition period when a new CMO commences work. Finding new CMOs or third-party suppliers involve additional cost and requires our management's time and focus. Any significant delay in the supply of ARCALYST or our product candidates or the raw materials needed to produce them, could considerably delay conducting our clinical trials and potential regulatory approval of our product candidates or our commercialization efforts. Our current and anticipated future dependence upon

others for the manufacture of ARCALYST and our product candidates may adversely affect our future profit margins and our ability to successfully commercialize any product candidates that receive marketing approval, if any, on a timely and competitive basis.

If we make manufacturing or formulation changes to our products or product candidates or change manufacturers or manufacturing processes, we may be unsuccessful in producing products or product candidates comparable to those used in prior clinical trials. Therefore, we may need to conduct additional process development or additional clinical trials to bridge our prior clinical results to those resulting from the new manufacturing process, which could impact the timing and subsequent success of our planned clinical trials. In addition, as we plan to produce clinical trial and commercial material at a CMO, the CMO may be required to adopt different manufacturing protocols or processes. For example, although Regeneron has produced ARCALYST for commercial use for over ten years, the FDA or other applicable regulatory authorities in other jurisdictions may reevaluate ARCALYST's current manufacturing processes or route of administration in connection with evaluating whether to approve ARCALYST for any new indication in the future or in connection with a technology transfer from Regeneron to us or another CMO.

The facilities used by our CMOs to manufacture ARCALYST and our current and future 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 or other regulatory authorities or based on their work for other clinical trial sponsors. 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 CMOs for compliance with cGMPs and other regulatory requirements in connection with the manufacture of ARCALYST and our product candidates. If our CMOs cannot successfully manufacture material that conforms to our specifications and the strict regulatory requirements of the FDA or other applicable regulatory authorities in other jurisdictions, they will not be able to secure or maintain regulatory approval for their manufacturing facilities. While we review the compliance history and performance of our CMOs and have the ability to audit their compliance and performance, we have no direct control over the ability of our CMOs to maintain adequate quality control, quality assurance and qualified personnel other than through quality monitoring in accordance with our agreements with the CMOs. If the FDA or comparable regulatory authorities in other jurisdictions 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 ARCALYST or our current or future 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 products or product candidates, if approved, operating restrictions and criminal prosecutions, any of which could significantly and adversely affect our business and supplies of our products or product candidates.

Although we have entered into certain agreements for the manufacture of clinical material for our product candidates and commercial material for ARCALYST, we may be unable to establish new agreements on acceptable terms, if at all, with third-party manufacturers for such product and 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 possibility that the third party cannot manufacture in quantities sufficient to meet our demand due to competing priorities, capacity limitations, or other reasons, including those related to the COVID-19 pandemic;
- 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. Furthermore, given the limited number of available manufacturing slots and the long lead times needed to reserve them, manufacturers require monetary commitments in connection with such reservations as well as fees for changes or cancellations in the reserved manufacturing slots. As a result, we may wait to reserve manufacturing slots until we can be informed by data from the clinical trials of our product candidates, which may be several months from the time we request manufacturing slots. Any significant delay in the supply of clinical materials for our product candidates could considerably delay conducting our clinical trials and potential regulatory approval of our product candidates. Alternatively, we may project when we may need additional clinical material for our product candidates and reserve manufacturing time-slots "at-risk" prior to our product candidates having generated data from their then current clinical trials. In addition, given the lead times we must provide to Regeneron with respect to the commercial supply of ARCALYST, we must place purchase orders based on projected demand, in advance of knowing the market acceptance of ARCALYST for the treatment of recurrent pericarditis. Such projections involve risks and uncertainties and may result in additional costs or delays in manufacturing clinical materials for ARCALYST and our product candidates when and if we actually need them and may result in having too little or too much ARCALYST in inventory to meet actual market demand.

Any performance failure on the part of our existing or future manufacturers could delay clinical development or marketing approval or commercialization efforts for our product candidates. 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. Further, Regeneron has an exclusive right to produce ARCALYST for a period of time, which could impact our ability to find a replacement manufacturer for ARCALYST in a short-period of time if needed.

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 ARCALYST or our 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' and suppliers' 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 and suppliers 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 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 our facility and the facilities of our third-party service providers, including as a result of the COVID-19 pandemic, could cause product shortages, disrupt or delay our clinical trials or regulatory approvals, delay or stop commercialization of our products and product candidates, and adversely affect our business.

The manufacture of our products and product candidates is highly regulated, complex and difficult, requiring a multi-step and controlled process, and even minor problems or deviations could result in the product candidates being out-of-spec, failed batches or other failures, such as defective products or manufacturing failures. We have limited experience overseeing the manufacturing processes of mavrilimumab, vixarelimab, and KPL-404 and no experience

overseeing the manufacturing process of ARCALYST. Due to the highly technical requirements of manufacturing our product candidates and the strict quality and control specifications, we and our third-party providers may be unable to manufacture or supply ARCALYST or 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, if any, and diminish our potential profitability, as applicable, which may lead to lawsuits or could delay the introduction of our product candidates to the market. Failure to produce sufficient quantities of ARCALYST could result in supply shortages for our patients, result in lost revenue, and impact our ability to hold sufficient quantities of safety stock to be properly positioned to address unexpected disruptions to the ARCALYST supply chain, which may lead to lawsuits.

The manufacture of ARCALYST and 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, failed batches and other supply disruptions. If microbial, viral or other contaminations are discovered in ARCALYST or 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, as well as disruptions in travel, shipping or delivery capabilities into and within the countries in which we or our manufacturers produce ARCALYST or our product candidates or disruptions to production capabilities, including due to the impact of natural disasters, accidents, boycotts, labor disputes, political and economic instability, including acts of terrorism or war, and an epidemic or pandemic or other outbreak of disease, including the COVID-19 pandemic. The occurrence of any such event could adversely affect our ability to satisfy the required supply for any of ARCALYST or our product candidates or successfully complete preclinical and clinical development, which would result in additional costs to us or impair our ability to generate revenue and would harm our business, financial condition and prospects significantly.

We and 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. If we or any of our third-party providers are not able to establish and maintain procedures and processes sufficient to satisfy cGMP standards, we could experience a delay, interruption or other issues in our manufacture, fill-finish, packaging, storage or delivery of ARCALYST or our product candidates, and any related 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, such as any impact due to the COVID-19 pandemic including shortages or reprioritizations of raw materials, reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, could result in a shortage of commercial products or product candidates or impose commercial product requirements, cause withdrawal of our product candidates or 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 drug substance and drug product used in ARCALYST and our product candidates are our sole source of supply, and the loss of any of these suppliers could significantly harm our business.

The drug substance and drug product used in ARCALYST, mavrilimumab and vixarelimab are supplied to us from single-source suppliers. Regeneron has a contractual right to be our sole source manufacturer of ARCALYST 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 drug substance and drug product 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 drug substance and drug product in the event any of our current suppliers of such drug substance and drug product 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 manufacturing ARCALYST, mavrilimumab and vixarelimab 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, alternative sources of commercial supply may need to be secured, 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 our manufacturing operations or one or more of our third-party manufacturers' or suppliers' relevant operations, such as due to the impact of the COVID-19 pandemic, including due to staffing shortages or reprioritizations, reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, production slowdowns or stoppages or interruptions in global shipping, the supply of the related product or product candidate or will be delayed until we or 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 preclinical and clinical programs or successfully commercialize our products could be materially and adversely impacted if any of the third-party suppliers upon which we rely for raw materials and preclinical and clinical stage product candidate and commercial stage product supply 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 manufacturing facilities or equipment or those of our third-party manufacturers' or suppliers' facilities or equipment may significantly impair our ability to manufacture our products and product candidates on a timely basis.

Establishing additional or replacement suppliers for the drug substance and drug product used in ARCALYST or our product candidates, if required, is unlikely to 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 or our CMOs 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 and our CMOs may seek to maintain adequate inventory of the drug substance and drug product used in ARCALYST or our product candidates, any interruption or delay in the supply of components or materials, or our inability to obtain such drug substance and drug product from alternate sources of comparable quality at acceptable prices in a timely manner could impede, delay, limit or prevent our development or commercialization efforts, which could harm our business, results of operations, financial condition and prospects.

Certain of the materials required in the manufacture and the formulation of our products and product candidates are derived from biological sources. Such 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 materials necessary for the manufacture of ARCALYST or 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 other material used in the manufacture of our products and product candidates could adversely impact or disrupt manufacturing, which would impair our ability to generate revenue from the sale of ARCALYST or our product candidates, if approved.

We rely, and expect to continue to rely, on third parties, including independent investigators and CROs, to activate sites, conduct and otherwise support our research activities, preclinical 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 rely on medical institutions, clinical investigators, contract laboratories and other third parties, such as CROs, to activate sites, conduct or otherwise support our preclinical studies and 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 such site activation, execution of and otherwise supporting clinical trials for our product candidates. While we have agreements governing their activities and we review the compliance history and performance of our CMOs as well as have the ability to audit such activities, we have no direct control over their activities and have limited influence over their actual performance other than through quality monitoring in accordance with our agreements with the CMOs. The third parties with whom we contract for execution of our preclinical studies and our clinical trials play a significant role in the conduct of these 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 preclinical studies and clinical trials in accordance with applicable GLP or GCP requirements, we 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 preclinical studies or clinical trials, we could be subject to warning letters or enforcement actions that may include civil penalties and criminal prosecution.

We and our CROs are 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, or EEA, and comparable regulatory authorities in other jurisdictions 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 regulatory authorities in other jurisdictions may require us to perform additional clinical trials before approving our marketing applications. We cannot ensure that, upon inspection, the FDA or comparable regulatory authorities in other jurisdictions 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 within certain timeframes. Failure to do so when required can result in fines, adverse publicity and civil and criminal sanctions.

Although we have and intend to continue to design the clinical trials for our product candidates, CROs will activate sites and conduct and oversee all of the clinical trials together with the various clinical trial sites that we engage to conduct the studies. As a result, many important aspects of our development programs for our product candidates, including their conduct and timing, will be outside of our direct control. Our reliance on third parties to activate sites and 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;
- have disruptions to their business and operations, including as a result of the impact from an epidemic or pandemic disease outbreak, including COVID-19 (see "Risk Factors — Risks related to product development — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact on our current or planned preclinical studies and clinical trials, which could be significant.");
- fail to comply with contractual obligations;
- have difficulty with or controlling the performance of their subcontractors;
- 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 activate sites and conduct and oversee our clinical trials and may subject us to unexpected cost increases that are beyond our control. If the CROs, their subcontractors or the clinical trial sites 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 may be materially and irreversibly harmed. If we are unable to rely on clinical data collected by our CROs, their subcontractors or the clinical trial sites, 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 CROs, their subcontractors or the clinical trial sites 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 preclinical studies or our clinical trials do not perform their contractual duties or obligations, experience significant business challenges, disruptions or failures, such as due to the impact of the COVID-19 pandemic, 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 preclinical 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 are suspended or 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, their subcontractors or the clinical trial sites 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 their failure to adhere to our clinical protocols, regulatory requirements or for other reasons, any

clinical trials such CROs, subcontractors or clinical trial sites 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, 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.

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, invention assignment agreements, material transfer agreements, collaborative research agreements, consulting agreements or other similar agreements with our collaborators, advisors, employees, independent contractors 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, Executing our Strategy, 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 or 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 product that modulates the signaling of IL-1α and IL-1β, anakinra (KINERET), marketed by Swedish Orphan Biovitrum AB, and one product that modulates the signaling of IL-1β, canakinumab (ILARIS), marketed by Novartis Pharmaceuticals Corporation. There are other therapies which modulates IL-1α in preclinical and clinical development for diseases other than recurrent pericarditis from Johnson & Johnson and XBIOTECH USA, INC.

We expect mavrilimumab, if further developed and approved for the treatment of GCA, to experience competitive pressure from tocilizumab (ACTEMRA), marketed by Genentech USA, Inc., which was approved in 2017 for use in GCA as an adjunct to steroid taper. Additional competition may be experienced from Eli Lilly and Company and AbbVie Inc., which are conducting clinical trials for oral janus kinase inhibitors, Sanofi S.A. and Regeneron, which are recruiting a Phase 3 clinical trial with their anti-IL-6 program, Novartis International AG, which is recruiting a trial with its IL-17 antagonist secukinumab (Cosentyx) and Janssen Biotech, Inc., which is testing ustekinumab (STELARA) in two small studies for GCA. There are multiple other programs targeting GM-CSF antagonism not currently pursuing GCA in clinical trials that could decide in the future to engage in development of therapies for GCA, including GlaxoSmithKline plc, Izana Bioscience, Roivant Sciences Ltd., I-Mab Biopharma Co., Ltd., and Humanigen, Inc.

We are also evaluating mavrilimumab for the treatment of COVID-19 pneumonia and hyperinflammation. There are currently hundreds of active, industry sponsored clinical trials testing many different mechanisms of action for the treatment of COVID-19 related therapeutic areas in addition to the approved vaccines, and the many other clinical trials testing vaccines for the prevention of COVID-19.

Multiple therapies are in development for prurigo nodularis, and any that receive FDA approval for this indication will be likely competitors to vixarelimab. These products include nemolizumab, dupilumab and nalbuphine ER. There are multiple agents targeting antagonism of the CD40/CD40L interaction across a variety of clinical uses including, Novartis International AG, Biogen Inc., or Biogen, and UCB, Inc., C.H. Boehringer Sohn AG & Ko. KG and AbbVie Inc., Eledon Pharmaceuticals, Inc, Annelixis Therapeutics LLC, ImmuNext Inc. and Sanofi S.A., Viela Bio, Bristol-Myers Squibb Company and Astellas Pharma Inc.

Further, the results of clinical trials for our product candidates may produce negative, inconclusive or uncompetitive results compared to those produced by any of these or other companies in the indications we are studying, which may result in us deciding, or regulators requiring us, to conduct additional clinical trials or modify or cease development programs for our product candidates. We may also determine that the potential product and commercial profile of any of our product candidates may not ultimately be commercially successful or even if they have the potential to ultimately be successful, we may not have sufficient recourses, which in either case could lead us to discontinue its development, or we may determine to not support further development of any of our product candidates at any time for any reason.

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, preclinical 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 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 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.

We may not be successful in executing our growth strategy to identify, discover, develop, in-license or acquire additional product candidates or technologies, and our growth strategy may not deliver the anticipated results or we may refine or otherwise alter our growth strategy. We may seek to acquire businesses or undertake business combinations, collaborations or other strategic transactions which may not be successful or on favorable terms, if at all, and we may not realize the intended benefits of such transactions.

We have acquired or in-licensed our existing product candidates, and as part of our strategy we plan to identify new product candidates or technologies that we believe are complementary to our existing product candidates. We may do this through our internal discovery program, or by acquiring the rights to product candidates and technologies through a variety of transaction types, including in-licensing, strategic transactions, mergers or acquisitions. If we are unable to identify, discover, develop, in-license or otherwise acquire and integrate product candidates, or their related companies, in accordance with this strategy, our ability to pursue this component of our growth strategy would be limited and we may need to refine or otherwise alter this strategy. We cannot be certain that we will be successful in such efforts, and

even if we are successful in such efforts, we cannot be certain that such discovery or transaction will be on favorable terms, or that, following any such discovery or transaction, we will be able to realize the intended benefits of it.

Research programs and business development efforts to identify new product candidates and technologies require substantial technical, financial and human resources. We may focus our efforts and resources on potential product candidates, technologies or businesses that ultimately prove to be unsuccessful. In-licensing and acquisitions of product candidates, technology or businesses often require significant payments and expenses and consume additional resources. We will need to continue to devote a substantial amount of time and personnel to research, develop and commercialize any such in-licensed or acquired product candidate or technology, or integrate any new business, and we may decide to reprioritize our efforts even after having expended resources on a particular prospect. Our research programs and business development efforts, including businesses or technology acquisitions, collaborations or licensing attempts, may fail to yield additional complementary or successful product candidates for clinical development and commercialization or successful business combinations 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 or businesses 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 or acquire businesses or undertake business combinations, collaborations, or other strategic transactions;
- for product candidates we seek to in-license or acquire or for businesses we seek to acquire or undertake business combinations, collaborations or other strategic transactions with, we may not be able to agree to acceptable terms with the licensor or owner of those product candidates or businesses;
- we may not succeed in formulation or process development;
- any product candidates to which we acquire the rights or that we discover may not succeed in preclinical studies or clinical trials or 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 any product candidates or technologies to which we acquire the rights or that we discover, obsolete or less attractive;
- any product candidates or technologies to which we acquire the rights may be covered by third-party patents or other exclusive rights;
- any product candidates or technologies to which we acquire the rights or that we discover may not allow us to leverage our expertise and our development and commercial infrastructure as currently expected;
- any product candidates or technologies to which we acquire the rights or that we discover will take substantial additional financial resources to develop and commercialize and we may not have sufficient funds to do so;
- the market for any product candidates or technologies to which we acquire the rights or that we discover may change during our program so that such a product or technology may become unreasonable to continue to develop;
- any product candidate to which we acquire the rights or that we discover may not be capable of being produced in commercial quantities at an acceptable cost, or at all; and

- any product candidate to which we acquire the rights or that we discover 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 to identify, discover, develop, in-license or acquire additional product candidates or technologies or to acquire businesses or undertake business combinations, collaborations, or other strategic transactions, or our growth strategy or strategic transactions may not deliver the anticipated results or we may refine or otherwise alter this strategy.

The consummation or performance of any acquisition, business combination, collaboration or other strategic transaction we may undertake in furtherance of our growth strategy or any refined or otherwise altered strategy, may involve additional risks, such as difficulties in assimilating different cultures, retaining personnel and integrating operations, which may be geographically dispersed, increased costs, exposure to liabilities, incurrence of indebtedness, or use a substantial portion of our available cash for all or a portion of the consideration or cause dilution to our existing shareholders if we issue equity securities for all or a portion of the consideration. If any of these events occurs or we are unable to meet our strategic objectives for any such transaction, we may not be able to achieve the expected benefits from the transaction and our business may be materially harmed.

We may seek to enter into collaboration, licensing or other strategic transactions or arrangements to further develop, commercialize or otherwise attempt to realize value from one or more of our product candidates, and any such transactions or arrangements that we may enter into may not be successful or be on favorable terms, which could adversely affect our ability to develop, commercialize or attempt to realize value from our product candidates.

We may seek to enter into collaboration, licensing or other strategic transactions or arrangements to further develop, commercialize or otherwise attempt to realize value from one or more of our product candidates depending on the merits of retaining rights to develop or commercialize the product candidates ourselves as compared to entering into such transactions or arrangements. In addition, we may seek to jointly develop, commercialize or otherwise exploit one or more of our product candidates with a third party. To the extent that we decide to enter into such transactions or arrangements, we will face significant competition in seeking appropriate collaborators, licensees or other strategic parties. Moreover, these transactions and arrangements are complex and time consuming to negotiate, document, implement and to close or maintain. We may not be successful in our efforts to establish collaborations, licenses or other strategic transactions or arrangements should we so chose to do so. The terms of any such transactions or arrangements that we may establish may have unfavorable tax consequences for our shareholders in the United States. 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 current or future collaborations, licenses or other strategic transactions or arrangements that we enter into may not be successful. The success of these potential collaboration, license arrangements and other strategic transactions or arrangements may depend heavily on the efforts and activities of our collaborators, sublicensees or other strategic parties. For example, in December 2019, we entered into a clinical collaboration with Kite to initiate a Phase 2 clinical trial evaluating the combination of Yescarta (axicabtagene ciloleucel) and mavrilimumab in relapsed or refractory large B-Cell lymphoma. Kite was to be the sponsor of this study and responsible for its conduct, but Kite later informed us that our clinical collaboration was discontinued due to a portfolio strategy review that impacted our trial as it had not started recruiting. Collaborations, licenses or other strategic transactions or arrangements are subject to numerous risks, which may include risks that the collaborator, licensee or other strategic party, as applicable:

- may have significant discretion in determining the efforts and resources that they will apply;
- may not commit sufficient resources to or otherwise not perform satisfactorily in carrying out its activities;
- 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;

- may own or co-own intellectual property covering products that results from our arrangement with them, and in such cases, we would not have the exclusive right to develop or commercialize such intellectual property, and even if we are able to license such exclusive rights, we may have to enter into a license agreement that include obligations to make milestone, royalty or other payments under such agreement; and

- may conduct sales and marketing activities or other operations that may not be in compliance with applicable laws, resulting in civil or criminal proceedings.

In addition, disputes may arise with respect to the ownership of any intellectual property developed pursuant to these arrangements. These arrangements may also be terminated, and, if terminated, may result in a need for additional capital to pursue further development or commercialization of the applicable product candidates.

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

We are highly dependent on the research and development, clinical, regulatory, manufacturing, commercial and business development expertise of members of our executive and senior management teams, as well as the other members of our management, scientific and clinical teams. Although we have entered into employment agreements with our executive officers and certain members of senior management, each of them or we may terminate their employment with us at any time. We do not maintain “key person” insurance for any of our executives, senior management 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 is also critical to our success. The failure to recruit, or the loss of the services of our executive officers, senior management or other key employees could impede the achievement of our research, development and commercialization objectives, including with respect to our sales, marketing and distribution capabilities, infrastructure and organization to commercialize products for which we have obtained marketing approval, including our lead program, ARCALYST in recurrent pericarditis, and for its other approved indications in the United States, which would seriously harm our ability to successfully implement our business strategy and potential commercial launch of ARCALYST. Furthermore, replacing executive officers, senior management 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. Changes in our senior management may be disruptive to our business, and, if we are unable to manage an orderly transition of responsibilities, our business may be adversely affected. 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 and clinical 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 need to continue to develop our company and expand our scope of operations, and we may encounter difficulties in managing this development and expansion, which could disrupt our operations.

We expect to continue to develop our company and expand 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 and infrastructure, expand our facilities over time and continue to recruit and train qualified personnel. Also, our executive and senior management teams have and may continue to divert a disproportionate amount of their attention away from their day-to-day activities and devote a substantial amount of time to managing these development

and expansion activities. For example in January 2021, we implemented select components of a new ERP system that will enable the organization to manage the complexity of operating a commercial organization more efficiently. As with any implementation this new system will require specific skills and expertise to setup, maintain and utilize the system. We may not be able to develop these skills internally or in sufficient time and capacity, which could require us to expend additional resources to acquire them. Due to our limited resources, certain employees have and may continue to perform activities that are beyond their regular scope of work, and we may not be able to effectively manage the development of our company, expansion of our operations or recruit and train qualified personnel. This may result in weaknesses of our systems and infrastructure, give rise to managerial, operational and financial mistakes, loss of business opportunities, loss of employees and reduced productivity among remaining employees. The development of our company and expansion of our operations may lead to significant costs and may divert financial resources from other projects, such as the development of one or more of our product candidates. If our executive and senior management teams are unable to effectively manage our anticipated 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 as planned, including with respect to our commercial launch of ARCALYST in recurrent pericarditis. Our future financial performance and our ability to commercialize our product candidates, if approved, and to compete effectively will depend, in part, on our ability to effectively manage the future development of our company and expansion of our operations.

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 vixarelimab. 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 and manufacture 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 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 Beth Israel Deaconess Medical Center to patent applications and patents related to KPL-404.

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, vixarelimab, KPL-404 or our other product candidates. In addition, the laws of other 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 EU 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 or are unable 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 in 2008, 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. Further, while patent term extension was awarded for relevant patents in certain European countries following the EMA's approval of ARCALYST for the treatment of CAPS, in 2012 the marketing authorization for CAPs was withdrawn. Patent term extensions may no longer be in effect or available, subject to the applicable laws in those countries as well as other factors, such as whether a marketing approval for ARCALYST is reissued and whether such reissuance is prior to the expiration of the patent's natural 20-year patent term. 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 in the United States covering ARCALYST as a composition of matter have expired, and patents covering ARCALYST as a composition of matter in Europe have a term that expires in 2023, not including any patent term extensions, and the patents covering mavrilimumab as a composition of matter have a term that expires in 2027 in the United States, not including any patent term adjustments (an adjustment to the term of the U.S. patent to compensate the patentee for delays caused by the USPTO during the examination process) or patent term extensions, and in 2027 in Europe, not including any patent term extensions. We may not receive any patent term extension for patents covering mavrilimumab as a composition of matter if such patent in an applicable jurisdiction expires before mavrilimumab would be eligible to receive regulatory approval in such jurisdiction. 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 ten years of marketing exclusivity in Europe. While we obtained orphan drug designations from the FDA for ARCALYST for the treatment of pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older, which includes the treatment of recurrent pericarditis, and for mavrilimumab for

the treatment of GCA, we may pursue orphan drug designation for our other product candidates in the United States and we may not be successful in obtaining such designation, or we may not be able to maintain the benefits of the designation for ARCALYST or mavrilimumab or any of our other product candidates. 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 the 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 “Risk Factors — Risks related to marketing approval and regulatory matters.”

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. Further, it is 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 other 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 or enforceable 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 or our licensors' former employees or consultants asserting an ownership right in our patents or patent applications, as a result of the work they performed on our or their behalf, respectively. 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 or our licensors 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 related to 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 or commercialize ARCALYST and our product candidates, mavrilimumab, vixarelimab 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 the MedImmune Agreement 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, or the Biogen Agreement, we acquired all of Biogen's right, title and interest in and to certain assets used in or relating to vixarelimab, including patents and other intellectual property rights, clinical data, know-how and inventory. In connection with our acquisition of Primatope Therapeutics, Inc., or Primatope, in March 2019, we acquired an exclusive world-wide license with Beth Israel Deaconess Medical Center for certain patent applications and patents related to KPL-404. 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. These agreements and any future such agreements that we enter into impose a variety of obligations and related consequences.

We are a party to 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 effect 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 elected to terminate the relevant agreement, which we have the right to do under each of these agreements. While we would expect to exercise our rights and remedies available to us in the event we fail to meet our obligations under these agreements in any material respect, 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. Termination of one of these agreements for any reason, and the related discontinuation of the development or commercialization of a product candidate could impair our ability to raise additional capital, generate revenue and may significantly harm our business, financial condition and prospects.

Regeneron has rights to develop ARCALYST in its retained fields of local administration to the eye and ear, oncology, deficiency of the IL-1 receptor, and CAPS, as well as for the treatment of Deficiency of the Interleukin-1 Receptor Antagonist, or DIRA, which was recently approved by the FDA. 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. The FDA approved ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021. Upon receipt of FDA approval for ARCALYST for the treatment of recurrent pericarditis, if any, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States and will evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. 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 the commercialization of ARCALYST for the treatment of 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 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 the oncostatin M receptor (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 Biogen 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 mavrilimumab and vixarelimab. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to mavrilimumab and vixarelimab 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 U.S. patent in court, we would need to overcome a statutory presumption of validity that attaches to every U.S. 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 or maintain the existing intellectual property rights we have, we may have to cease 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 patent lawsuit outside of the United States, alleging our infringement of a competitor's patents, we could be prevented from marketing our products in one or more such 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 governmental patent agencies outside of the United States 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 patent agencies outside of the United States 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 intellectual property laws outside of the United States. In addition, the patent laws of some such 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 jurisdictions outside of the United States. 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 countries outside of the United States 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 jurisdictions outside of the United States, 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 other 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 our patents that may be asserted against us by our competitors or other third parties. Any of these outcomes could have a material adverse effect on our business.

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, independent contractors and consultants, and invention assignment agreements with our independent contractors, 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. 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 in the United States or jurisdictions outside of the United States, 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 product candidates in the United States or jurisdictions outside of the United States 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 product candidates in the United States or any jurisdiction outside of the United States. 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 jurisdictions outside of the United States, 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.

Other Risks Related to Our Business

The COVID-19 pandemic, and measures taken in response to the pandemic or the easing of such measures, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position.

The COVID-19 pandemic, and measures taken in response to the pandemic or the easing of such measures, could cause significant disruption in our business and operations and could cause significant disruption the business and operations of our manufacturers, CROs upon whom we rely on to conduct our clinical trials, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities.

The federal and state governments in the United States and the governments of other countries around the globe have implemented various measures in response to the COVID-19 pandemic, including significant restrictions on businesses as well as travel into and within the countries in which our manufacturers produce our product candidates or where we conduct our clinical trials or otherwise conduct business or engage with other third parties. In response to the COVID-19 pandemic and measures introduced by state and federal governments in the United States, we implemented workplace protocols at our facilities. While the majority of our employees are able to carry out their responsibilities working outside of our physical locations, for our essential workers and those choosing to return to our offices to carry out their responsibilities, we implemented additional safety measures, including occupancy limits, restricting business travel, providing and requiring the use of personal protective equipment, self-screening prior to accessing our facilities, and others. We continue to monitor the developments, restrictions and requirements in jurisdictions where we have offices, and plan to update the protocols for our offices as applicable. If the COVID-19 pandemic and measures undertaken in response to the pandemic are prolonged, or the easing of any of such measures has significant adverse consequences, we may experience and our manufacturers, CROs or other third parties with whom we conduct business or otherwise engage, may experience or continue to experience staffing shortages or reprioritizations, production slowdowns or stoppages, and disruptions in delivery systems now or in the future. For example, the COVID-19 pandemic and measures taken in response to the pandemic, including business and travel restrictions and social-distancing to halt the spread of the pandemic, has had an impact on certain aspects of our commercialization strategy, including interacting with third-party payors, physicians and patient advocacy groups to build disease awareness, and conducting in-person market research as well as recruiting qualified candidates to enhance our commercial operations and support commercialization, which, if prolonged, may impede the effective commercialization of our product candidates and result in lower than anticipated future revenue.

The COVID-19 pandemic may also have a significant adverse impact our preclinical studies and clinical trials, which could significantly impede, delay, limit or prevent the clinical development of our product candidates and ultimately lead to the delay or denial of regulatory approval of our product candidates, which would materially adversely affect our business and operations, including our ability to generate revenue. See “Risk Factors — Risks related to product development — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact our current or planned preclinical studies and clinical trials, which could be significant.”

Moreover, the COVID-19 pandemic is impacting the global economy, and the U.S. economy in particular, with the potential for the economic downturn to be severe and prolonged. A severe or prolonged economic downturn could

result in a variety of risks to our business, including disruptions in the financial markets. For example, the trading prices of biopharmaceutical companies have been highly volatile as a result of the COVID-19 pandemic. These disruptions could adversely impact our ability to raise additional capital when needed or on acceptable terms, if at all.

The COVID-19 pandemic and measures undertaken in response to the pandemic continue to rapidly evolve. There is uncertainty relating to the potential effect of COVID-19 on our business and operations. The extent of the impact on our business and operations will depend on future developments, which are highly uncertain and cannot be predicted with confidence, such as the ultimate geographic spread of the disease, the duration of the pandemic, business and travel restrictions, quarantines, shelter-in-place orders and social distancing in the United States and other countries, business closures or business disruptions, the availability and efficacy of vaccines, the effectiveness of other actions taken in the United States and other countries to contain and treat the disease, and the impact of any easing of such measures.

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 face an inherent risk of product liability exposure related to the testing of our product candidates in human clinical trials and the commercialization of ARCALYST and any product candidates that we may develop, if approved. 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. 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.

The United Kingdom's withdrawal from the European Union may have a negative effect on global economic conditions, financial markets and our business.

Following a national referendum and enactment of legislation by the government of the United Kingdom, the United Kingdom formally withdrew from the European Union and ratified a trade and cooperation agreement governing its future relationship with the European Union. The agreement, which was being applied provisionally from January 1, 2021 and was ratified by the European Parliament and the Council of the European Union in April 2021, addresses trade,

economic arrangements, law enforcement, judicial cooperation and a governance framework including procedures for dispute resolution, among other things. Because the agreement merely sets forth a framework in many respects and will require complex additional bilateral negotiations between the United Kingdom and the European Union as both parties continue to work on the rules for implementation, significant political and economic uncertainty remains about how the precise terms of the relationship between the parties will differ from the terms before withdrawal.

These developments, or the perception that any related developments could occur, have had and may continue to have a material adverse effect on global economic conditions and the financial markets, and may significantly reduce global market liquidity, restrict the ability of key market participants to operate in certain financial markets or restrict our access to capital. Any of these factors could have a material adverse effect on our business, financial condition and results of operations and reduce the price of our shares.

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

In the United States, EU 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 operations. For example, in the United States, the Affordable Care Act substantially changed 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, 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, executive and Congressional challenges to certain aspects of the Affordable Care Act. For example, 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. On December 14, 2018, a U.S. District Court Judge in Texas ruled that the Affordable Care Act is unconstitutional in its entirety because the “individual mandate” was repealed by Congress as part of the Tax Act. On December 18, 2019, the U.S. Court of Appeals for the 5th Circuit upheld the district court’s decision that the individual mandate was unconstitutional but remanded the case back to the District Court to determine whether the remaining provisions of the Affordable Care Act are invalid as well. The U.S. Supreme Court is currently reviewing the case, although it is unclear when a decision will be made or how the Supreme Court will rule. In addition, there may be other efforts to challenge, repeal or replace the ACA, or portions thereof, which will affect our business. It is possible that the ACA, as currently enacted or as it may be amended in the future, and other healthcare reform measures that may be adopted in the future, could have a material adverse effect on our industry generally and on our ability to successfully commercialize our product candidates, if approved.

In addition, other legislative changes have been proposed and adopted in the United States since the Affordable Care Act was enacted. 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 2030, with the exception of a temporary suspension from May 1, 2020 through December 31, 2021, unless additional Congressional action is taken. The American Taxpayer Relief Act of 2012, 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 Congressional inquiries and proposed and enacted 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. For example, in September 2018, CMS announced that it will allow Medicare Advantage Plans the option to use step therapy for Part B drugs beginning January 1, 2019. 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 and municipalities 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, if approved, or put pressure on our product pricing.

In the EU, 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 EU, 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 health care 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 the EU, 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 EU or elsewhere. For example, the new presidential administration may change governmental policies and regulations that affect our operations and business, including our clinical trials, regulatory approval, pharmaceutical pricing and reimbursement. 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 or operational 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 COVID-19 pandemic is impacting the global economy with the potential for the economic downturn to be severe and prolonged. A severe or prolonged economic downturn caused by the economic impact from the COVID-19 pandemic could result in a variety of risks to our business, such as disruptions to our operations and the operations of our manufacturers, CROs or other third parties with whom we conduct business or engage, including as a result of disruptions in travel into and within the countries in which we conduct our clinical trials or our manufacturers produce our product candidates or we conduct business or otherwise engage with such other third parties. These disruptions could adversely affect our ability to satisfy the required supply for any of our product candidates or successfully complete preclinical and clinical development of our product candidates, which could require us to incur additional costs, and impair our ability to obtain regulatory approval of our product candidates and generate

revenue. A severe or prolonged economic downturn could also impair our ability to raise additional capital when needed or on acceptable terms, if at all. Doing business internationally involves a number of other 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 operations outside of the United States;
- 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 technology systems, or those of our third-party CMOs, CROs or other contractors, consultants and service providers, may fail or suffer cyber-attacks or security breaches, which could result in a material disruption of our or such third-party's business or operations and our development programs for our product candidates' or loss of other assets, including funds.

Despite the implementation of security measures and cyber-security insurance, our internal technology systems and those of our third-party CMOs, CROs and other contractors, consultants and service providers as well as employees that are working outside of our facilities are vulnerable to damage from viruses, unauthorized access and attacks, theft, natural disasters, terrorism, war and telecommunication and electrical failures. As a result of the COVID-19 pandemic, we may experience increased cybersecurity risks due to the impacts from prolonged remote work arrangements. Attacks upon information technology systems are increasing in their frequency, levels of persistence, sophistication and intensity, and are being conducted by sophisticated and organized groups and individuals with a wide range of motives and expertise. As a result of the COVID-19 pandemic, we may also face increased cybersecurity risks due to our reliance

on internet technology and the number of our employees who are working remotely, which may create additional opportunities for cybercriminals to exploit vulnerabilities. Furthermore, because the techniques used to obtain unauthorized access to, or to sabotage, systems change frequently and often are not recognized until launched against a target, we may be unable to anticipate these techniques or implement adequate preventative measures. We may also experience security breaches that may remain undetected for an extended period. If such an event were to occur and cause interruptions in our business and operations or those of our third-party CMOs, CROs and other contractors, consultants and service providers as well as employees that are working outside of our facilities, it could result in a material disruption of our or such third-party's business or operations and our development programs of our product candidates' or loss of other assets, including funds. 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 not covered by our cyber-security insurance and the further development of our product candidates could be delayed.

Actual or perceived failures to comply with applicable data protection, privacy and security laws, regulations, standards and other requirements could adversely affect our business, results of operations, and financial condition.

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. We are not currently acting as a covered entity or business associate under HIPAA and thus are not subject to its requirements or penalties. However, any person may be prosecuted under HIPAA's 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 HIPAA's 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.

In addition, certain state laws govern the privacy and security of health information in certain circumstances, some of which are more stringent than HIPAA and many of which differ from each other in significant ways and may not have the same effect, thus complicating compliance efforts. Failure to comply with these laws, where applicable, can result in the imposition of significant civil and/or criminal penalties and private litigation. For example, California enacted the California Consumer Privacy Act, or the CCPA, on June 28, 2018, which went into effect on January 1, 2020. The CCPA gives California residents expanded rights to access and delete their personal information, opt out of certain personal information sharing and receive detailed information about how their personal information is used. The CCPA provides for civil penalties for violations, as well as a private right of action for data breaches that is expected to increase data breach litigation. The CCPA may increase our compliance costs and potential liability. Further, the California Privacy Rights Act, or CPRA, recently passed in California. The CPRA will impose additional data protection obligations on covered businesses, including additional consumer rights processes, limitations on data uses, new audit requirements for higher risk data, and opt outs for certain uses of sensitive data. It will also create a new California data protection agency authorized to issue substantive regulations and could result in increased privacy and information security enforcement. The majority of the provisions will go into effect on January 1, 2023, and additional compliance investment and potential business process changes may be required.

Our clinical trial programs outside the United States may implicate international data protection laws, including the EU General Data Protection Regulation and legislation of the EU member states implementing it, or GDPR, and legislation of the EU and EEA member states implementing it. The GDPR went into effect in May 2018 and imposes strict requirements for processing the personal data of individuals within the EEA. Companies that must comply with the GDPR face increased compliance obligations and risk, including more robust regulatory enforcement of data protection requirements and potential fines for noncompliance of up to €20 million or 4% of the annual global revenues of the noncompliant company, whichever is greater. Further from January 1, 2021, we may be subject to the GDPR and also

the UK GDPR, which, together with the amended UK Data Protection Act 2018, retains the GDPR in UK national law. The UK GDPR mirrors the fines under the GDPR, e.g. fines up to the greater of €20 million (£17.5 million) or 4% of global turnover. The relationship between the United Kingdom and the European Union in relation to certain aspects of data protection law remains unclear, and it is unclear how UK data protection laws and regulations will develop in the medium to longer term, and how data transfers to and from the United Kingdom will be regulated in the long term. These changes will lead to additional costs and increase our overall risk exposure. Currently there is a four to six-month grace period agreed in the EU and UK Trade and Cooperation Agreement, ending June 30, 2021 at the latest, whilst the parties discuss an adequacy decision. The European Commission published a draft adequacy decision on February 19, 2021. If adopted, the decision will enable data transfers from EU member states to the United Kingdom for a four-year period, subject to subsequent extensions. Failure by our CROs and other third-party contractors to comply with the strict rules on the transfer of personal data outside of the EU 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 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 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.

Securities class action and derivative lawsuits and other legal proceedings are often brought against companies, which could result in substantial costs and divert management's attention.

Securities class action and derivative lawsuits and other legal proceedings are often brought against companies following a decline in the market price of their securities. There can be significant fluctuations in market price for the securities of early-stage biotechnology companies, such as us. As a result, we may be more susceptible to these types of lawsuits and legal proceedings than other companies with more stable security prices. In connection with any litigation or other legal proceedings, we could incur substantial costs, and such costs and any related settlements or judgments may not be covered by insurance. We could also suffer an adverse impact on our reputation and a diversion of management's attention and resources, which could have a material adverse effect on our business.

Although we maintain director and officer liability insurance coverage, it may not be adequate to cover all liabilities that we may incur and is subject to deductibles and coverage limitations. 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 class action and derivative lawsuits and other legal proceedings or claims often brought against companies following a decline in the market price of their securities, we will be exposed to significant liabilities, which may materially and adversely affect our business and financial position.

We and our employees and third parties with whom we contract are increasingly utilizing social media tools as a means of communication both internally and externally, and noncompliance with applicable requirements, policies or contracts due to social media use or negative posts or comments could have an adverse effect on our business.

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 products, product

candidates or business may cause us to be found in violation of applicable requirements. In addition, our employees or third parties with whom we contract, such as our CROs or CMOs, 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 or information regarding our product candidates or clinical trials. Clinical trial patients may also knowingly or inadvertently make use of social media in ways that may not comply with legal or contractual requirements for participation in a clinical trial, including with respect to any AEs they may experience, which may give rise to liability and regulatory risk. Furthermore, negative posts or comments about us or our products, 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 noncompliance 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 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 preclinical studies or clinical trials, which could result in regulatory sanctions and cause serious harm to our reputation.

It is not always possible to identify and deter misconduct by employees and other third parties. 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.

Risks Related to Our Common Shares

The concentration of ownership of our Class B common shares, which are held primarily by our executive officers and certain other members of our senior management, and the conversion rights of the holders of our Class A1 common shares, which shares are held primarily by entities affiliated with certain of our directors, and Class B1 common shares, all of which shares are held by entities affiliated with certain of our directors means that such persons are, and such entities may in the future be, able to influence certain matters submitted to our shareholders for approval; and such concentration and conversion rights and resulting concentration of control may have an adverse effect on the price of our Class A common shares and may result in our Class A common shares being undervalued.

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. Our Class A1 common shares and Class B1 common shares have no voting rights. As a result, all matters submitted to our shareholders are decided by the vote of holders of our Class A common shares and Class B common shares. As a result of the multi-class voting structure of our common shares, the holders of our Class B common shares, which consist primarily of our executive officers and certain other members of our senior management, collectively control over a majority of the combined voting power of our common shares and therefore are able to control the outcome of certain matters submitted to our shareholders for approval. As of March 31, 2021, the holders of Class A common shares accounted for approximately 63% of our aggregate voting power and the holders of Class B common shares accounted for approximately 37% of our aggregate voting power. Our executive officers and certain other members of our senior management hold Class A common shares and Class B common shares representing approximately 34% of our aggregate voting power as of March 31, 2021 and may have the ability to influence the outcome of certain matters submitted to our shareholders for approval.

However, this percentage may change depending on any conversion of our Class B common shares, Class A1 common shares or Class B1 common shares. Each holder of our Class B common shares has the ability to convert any portion of its Class B common shares into Class B1 common shares or Class A common shares at any time with advance notice to us. Each holder of our 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 with advance notice to us, 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 with advance notice to us. Our Class A1 common shares and Class B1 common shares cannot be converted if, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of our issued and outstanding Class A common shares unless such holder provides us with 61-days' prior notice that it intends to increase, decrease or waive such threshold upon conversion. As of March 31, 2021, entities affiliated with certain members of our directors could convert their Class A1 common shares and Class B1 common shares upon 61-days' prior written notice into Class A common shares and Class B common shares, respectively, which in the aggregate would result in such entities holding approximately 78% of our aggregate voting power and having the ability to control the outcome of certain matters submitted to our shareholders for approval.

Due to these conversion rights, holders of our Class A1 common shares and our Class B1 common shares could, at any time with appropriate advance notice to us, 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 and would decrease the ability of the current holders of our Class A common shares and Class B common shares to influence or control matters submitted to our shareholders for approval. In addition, the conversion of Class B common shares to Class A or Class B1 common shares will have the effect of increasing the relative voting power of the holders of Class B common shares who retain their shares in the long term.

These conversion rights as well as concentrated control that limit certain shareholders' ability to influence corporate matters may have an adverse effect on the price of our Class A common shares, including our Class A common shares being undervalued. Holders of our Class B common shares collectively control our management and affairs and are able to influence or control the outcome of certain matters submitted to our shareholders for approval, including the election of directors. Due to the conversion rights of the holders of our Class A1 and B1 common shares, entities affiliated with certain of our directors could 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. As of April 30,

2021, entities affiliated with certain of our directors held 71% of our Class A1 common shares and 100% of our Class B1 common shares. Upon 61-days' prior written notice, these entities could convert their Class A1 common shares and Class B1 common shares into Class A common shares and Class B common shares, which in the aggregate would result in such entities holding approximately 78% of the voting power of our outstanding share capital. In addition, this concentration of control might adversely affect certain corporate actions that some of our shareholders may view as beneficial, for example, 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.

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

Our share price is likely to continue to be volatile. The stock market in general and the market for biopharmaceutical companies in particular have experienced extreme volatility, including as a result of the COVID-19 pandemic, that has often been unrelated to the operating performance of particular companies. As a result of this volatility, our shareholders may not be able to sell their Class A common shares at or above the price they paid for their shares. 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 the commencement, enrollment and the ultimate completion of clinical trials;
- delays in approvals of our product candidates from the PDUFA goal date or failures in obtaining approval of our product candidates;
- the results and potential impact of competitive products or technologies;
- our ability to manufacture and successfully produce our product candidates;
- our ability to commercialize our product candidates, if approved;
- the size of the market for our product candidates;
- actual or anticipated changes in estimates as to financial results, capitalization, 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 our inability to obtain additional funding;
- failure to meet or exceed the 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 or from our entering into entering collaborations or other strategic transaction agreements;
- changes in the structure of healthcare payment systems;
- market conditions in the pharmaceutical and biotechnology sectors;
- general economic, industry and market conditions, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic;
- changes in voting control of, or sales of our shares by, our executive officers and certain other members of our senior management or entities affiliated with certain of our directors that hold our shares; and
- the other factors described in this “Risk Factors” section.

In addition, given the limited trading history of our Class A common shares, there is a risk that an active trading market for our shares will not be sustained, which could put downward pressure on the market price of our common shares and thereby affect the ability of our shareholders to sell their shares. An inactive trading market may also impair our ability to raise capital to continue to fund operations by selling shares and may impair our ability to acquire other companies or technologies by using our shares as consideration.

Additionally, the trading prices of biopharmaceutical companies have been highly volatile as a result of the COVID-19 pandemic. The COVID-19 outbreak continues to rapidly evolve. The extent to which the outbreak may impact our business in the future, including our preclinical studies and clinical trials, results of operations and financial condition will depend on future developments, which are highly uncertain and cannot be predicted with confidence. Such factors include but are not limited to the ultimate geographic spread of the disease, the duration of the pandemic, business and travel restrictions, quarantines, shelter-in-place orders and social distancing in the United States and other countries, business closures or business disruptions, the availability and efficacy of vaccines, the effectiveness of other actions taken in the United States and other countries to contain and treat the disease and the impact of any easing of such measures.

If securities or industry analysts cease publishing 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 is influenced by the research and reports that equity research analysts publish about us and our business. We do not have any control over the analysts or the content and opinions included in their reports. The price of our Class A common 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 number of our Class A common shares in the public market, including Class A common shares issuable upon conversion of our Class B, Class A1 and Class B1 common shares, could cause the share price of our Class A common shares to fall.

A significant number of our Class A common shares are issuable upon conversion of our Class B, Class A1, and Class B1 common shares. Our Class B and Class B1 common shares automatically convert into Class A common shares upon transfer by a holder of such shares to persons or entities not affiliated with such holder. In addition, each holder of our Class B common shares has the ability to convert any portion of its Class B common shares into Class B1 common shares or Class A common shares at any time with advance notice to us, each holder of our 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 with advance notice to us 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 with advance notice to us. However, our Class A1 common shares and Class B1 common shares cannot be converted if, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of our issued and outstanding Class A common shares unless such holder provides us with 61-days' prior notice that it intends to increase, decrease or waive such threshold upon conversion.

As of March 31, 2021 upon conversions of our Class B, Class A1, and Class B1 common shares, approximately 2.3 million of additional Class A common shares were issuable and eligible for resale in the public market to the extent permitted by the provisions of Rule 144 promulgated under the Securities Act of 1933, as amended, or the Securities Act, and such rule, Rule 144. In addition, as of March 31, 2021, there were approximately 10.9 million Class A common shares subject to outstanding share options and restricted share units under our equity incentive plans that may become eligible for sale in the public market to the extent permitted by the provisions of applicable vesting schedules and Rule 144 and Rule 701 under the Securities Act.

Over a majority of our common shares are held by our executive officers and other members of our senior management together with entities affiliated with certain of our directors. As of March 31, 2021, on an as-converted to Class A common shares basis, these shareholders collectively held approximately 34.4 million of our Class A common shares. If any of these shareholders sell, convert or transfer, or indicate an intention to sell, convert or transfer, a substantial amount of their common shares (after certain restrictions on conversion or resale lapse), the market price of our Class A common shares could decline.

Pursuant to our amended and restated investor rights agreement, or our investors rights agreement, certain shareholders are entitled to certain registration rights with respect our Class A common shares, including Class A common shares issuable upon conversions of our Class B, Class A1, and Class B1 common shares and upon the exercise of certain rights to acquire Class A common shares, or collectively registerable securities, under the Securities Act. As of March 31, 2021, on an as-converted to Class A common shares basis, we have registered approximately 31.8 million Class A common shares held by certain holders affiliated with certain of our directors as well as certain other shareholders pursuant to our investor rights agreement, which are freely tradable without restriction under the Securities Act, to the extent permitted by Rule 144. Further, pursuant to the investors rights agreement (a) the holders affiliated with certain of our directors are entitled to certain registration rights under the Securities Act with respect to registrable securities they may own now or in the future and (b) our executive officers are also entitled to certain registration rights under the Securities Act with respect to registrable securities they may own now or in the future, including, on an as-converted to Class A common shares basis, the approximately 1.6 million Class A common shares held by our executive officers as of March 31, 2021. If any of these 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.

Future sales or issuances of our common shares or rights to purchase common shares, including under our shelf registration statement or 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 additional Class A common shares, Class B common shares, Class A1 common shares, Class B1 common shares or other 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 under our shelf registration statement or otherwise. 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.

In addition, the consummation or performance of any acquisition, business combination, collaboration or other strategic transaction we may undertake in furtherance of our growth strategy may cause dilution to our existing shareholders if we issue equity securities for all or a portion of the consideration.

We will continue to incur increased costs as a result of operating as a public company, including in connection with becoming a large accelerated filer, and our management will be required to devote substantial time to new compliance initiatives.

As a public company, we incur significant legal, accounting and other expenses. In addition, the Sarbanes-Oxley Act of 2002 and rules subsequently implemented by the SEC and The Nasdaq Global Select Market, or Nasdaq, where our Class A common shares are listed, impose various requirements on public companies, including establishment and maintenance of effective disclosure and financial controls and corporate governance practices. Our management and other personnel devote a substantial amount of time to these compliance initiatives. Moreover, these rules and regulations increase our legal and financial compliance costs and make some activities more time-consuming and costly, which we expect to increase in connection with our being a large accelerated filer and no longer qualifying as an emerging growth company and smaller reporting company. In addition, we expect our costs to increase as we comply with requirements that we were previously exempt from as an emerging growth company and smaller reporting company and cause management and other personnel to divert attention from operational and other business matters to devote substantial time to public company reporting requirements.

Pursuant to Section 404, we are 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. To maintain compliance with Section 404, we engage 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, engage outside consultants and refine and revise 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 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 contain provisions that could make it more difficult for a third party to acquire us. These provisions provide for:

- a classified board of directors with staggered three-year terms;
- directors only to be removed for cause;
- an affirmative vote of 66 2/3% of the voting power of our voting shares for certain “business combination” transactions that have not been approved by our board of directors;
- our multi-class common share structure, which provides our holders of Class B common shares with the ability to significantly influence the outcome of matters requiring shareholder approval, even if they own less than a majority of our outstanding Class A common shares;
- 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 our shareholders to elect directors of their choosing and cause us to take corporate actions other than those our shareholders desire.

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

We have never declared or paid cash dividends on our 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 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 the sole source of gain for our shareholders 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 our shareholders 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 amended and restated 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.

Our amended and restated bye-laws designate the Supreme Court of Bermuda as the choice of jurisdiction for disputes that arise concerning the Bermuda Companies Act 1981, as amended, or the Companies Act, or out of or in connection with our amended and restated bye-laws, which could limit our shareholders' ability to choose the judicial forum for disputes with us or our directors or officers.

Our amended and restated bye-laws provide that, unless we consent in writing to the selection of an alternative jurisdiction, any dispute that arises concerning the Companies Act, or out of or in connection with our amended and restated bye-laws, including any question regarding the existence and scope of any bye-law or whether there has been a breach of the Companies Act or the amended and restated bye-laws by any of our officers or directors (whether or not such a claim is brought in the name of a shareholder or in the name of our company) shall be subject to the jurisdiction of the Supreme Court of Bermuda.

Any person or entity purchasing or otherwise acquiring any interest in any of our shares shall be deemed to have notice of and consented to this provision. This choice of jurisdiction provision may limit a shareholder's ability to bring a claim in a judicial forum of its choosing for disputes with us or our directors or officers, which may discourage lawsuits against us and our directors and officers. If a court were to find either choice of jurisdiction provision in our amended and restated bye-laws to be inapplicable or unenforceable in an action, we may incur additional costs associated with resolving the dispute in other jurisdictions, which could harm our results of operations.

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 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 amended and restated 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 Nasdaq. This general permission would cease to apply if we were to cease to be listed on Nasdaq.

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 other 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 subsidiaries in the United States, the United Kingdom, Germany, Switzerland and France. If we succeed in growing our business, we expect to conduct increased operations through our subsidiaries in various tax jurisdictions subject to transfer pricing arrangements between us and such 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 laws related to tax practices and substance requirements in Bermuda and other jurisdictions could adversely affect our operations.

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 were 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 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;
- changes to and 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.

Bermuda enacted the Economic Substance Act 2018, which was amended as recently as December 24, 2019 and issued Economic Substance Regulations in 2018, which were amended as recently as December 24, 2019 (collectively, “ES Laws”). Pursuant to the ES Laws, certain entities in Bermuda engaged in “relevant activities” are required to maintain appropriate physical presence in Bermuda and to satisfy economic substance requirements commencing as of January 1, 2019, with a six-month transition period until July 1, 2019. The list of “relevant activities” includes carrying on as a business in any one or more of the following categories: banking, insurance, fund management, financing, leasing, headquarters, shipping, distribution and service center, intellectual property and holding entities. Under the ES Laws, any relevant entity must satisfy economic substance requirements locally or face financial penalties, restriction or regulation of its business activities or may be struck off as a registered entity from the Bermuda Registrar of Companies. Because we do not report gross revenue attributable to any such relevant activity, under Section 6 of the Revised Final Guidance Notes issued on September 18, 2020, we believe that we are not obliged to meet the economic substance requirements. We will continue to monitor our status with respect to the ES Laws based on our results of operations, and whether further action may be required in the future by the Company to be in compliance with the ES Laws.

While we believe we are not a passive foreign investment company (“PFIC”) for U.S. federal income tax purposes for the year ended December 31, 2020 and do not believe we will be a PFIC for the year ended 2021, if we were to be

classified a PFIC this could result in adverse U.S. federal income tax consequences to U.S. investors in our common shares.

We recently completed an analysis of the Company's and its subsidiaries sources of income and character of their assets for U.S. federal income tax purposes and determined that neither the Company nor any of its subsidiaries would be classified a PFIC for the taxable year ended December 31, 2020. However, the U.S. tax authorities could disagree with our analysis and consider the Company, or our subsidiaries, to be a PFIC for the 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 (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, or our subsidiaries, are classified as a PFIC in any year with respect to which a U.S. Holder owns our 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 PFIC test described above, unless we cease to be a PFIC and the U.S. Holder made a "qualified electing fund" election or "mark-to-market" election for (i) the first taxable year the U.S. Holder was treated as owning our shares while we were a PFIC or (ii) for the taxable year in which we were a PFIC and the U.S. Holder made a "deemed sale" election or was qualified to and made a "deemed dividend" election. A "U.S. Holder" is a beneficial owner of our 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 (i) is subject to the supervision of a U.S. court and all substantial decisions of which are subject to the control of one or more "United States persons" (within the meaning of Section 7701(a)(30) of the U.S. Internal Revenue Code of 1986, as amended), or (ii) has a valid election in effect to be treated as a United States person for U.S. federal income tax purposes.

If we are classified as a PFIC for any taxable year during which a U.S. Holder holds our Class A common shares, certain adverse U.S. federal income tax consequences could apply to such U.S. Holder, including (i) the treatment as ordinary income of any gain realized on a disposition of our shares and distributions on our shares not being qualified dividend income, (ii) the application of a deferred interest charge on the tax on such gain and distributions, and (iii) the obligation to comply with certain reporting requirements.

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

We believe we will likely be classified as a controlled foreign corporation for the taxable year ended December 31, 2021. Even if we were not classified as a controlled foreign corporation, certain of our non-U.S. subsidiaries could be treated as controlled foreign corporations because our group includes one or more U.S. subsidiaries. If a U.S. Holder is treated as owning (directly, indirectly or constructively) at least 10% of the value or voting power of our 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," or GILTI, and investments in U.S. property by such controlled foreign corporation, regardless of whether such corporation makes 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 or income inclusions may subject such shareholder to significant monetary penalties and may prevent the statute of limitations with respect to such shareholder's 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 such investor is treated as a United States shareholder with respect to us or any of our non-U.S. subsidiaries. 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 any investment in our Class A common shares.

Item 2. Unregistered Sales of Equity Securities and Use of Proceeds

Issuance of Unregistered Equity Securities

None.

Item 3. Defaults Upon Senior Securities.

None.

Item 4. Mine Safety Disclosures.

None.

Item 5. Other Information.

None.

Item 6. Exhibits

Exhibit Number	Exhibit Description	Incorporated by Reference				Filed/ Furnished Herewith
		Form	File No.	Exhibit	Filing Date	
3.1	Memorandum of Association of Kiniksa Pharmaceuticals, Ltd.	S-1	333-224488	3.1	4/27/2018	
3.2	Amended and Restated Bye-Laws of Kiniksa Pharmaceuticals, Ltd.	8-K	001-38492	3.1	5/29/2018	
4.1	Specimen Share Certificate evidencing the Class A common shares	S-1/A	333-224488	4.1	5/14/2018	
4.2	Second Amended and Restated Investors' Rights Agreement, dated as of February 9, 2018	S-1	333-224488	3.1	4/27/2018	
10.1†	Commercial Supply Agreement, dated February 26, 2021, by and between Kiniksa Pharmaceuticals (UK) Ltd. and Regeneron Pharmaceuticals, Inc.					*
10.2†	Amendments Nos. 1 and 2 to the License Agreement, dated September 25, 2017, by and between Kiniksa Pharmaceuticals and Regeneron Pharmaceuticals, Inc.					*
10.3	Employment Agreement, effective as of April 1, 2021, by and between the Company and Mark Ragosa					*
10.4	Non-Employee Director Compensation Program					*
31.1	Rule 13a-14(a) / 15d-14(a) Certification of Chief Executive Officer					*
31.2	Rule 13a-14(a) / 15d-14(a) Certification of Chief Financial Officer					*
32.1	Section 1350 Certification of Chief Executive Officer					**
32.2	Section 1350 Certification of Chief Financial Officer					**
101.INS	XBRL Instance Document – the instance document does not appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL document					***

Exhibit Number	Exhibit Description	Incorporated by Reference				Filed/ Furnished Herewith
		Form	File No.	Exhibit	Filing Date	
101.SCH	Inline XBRL Taxonomy Extension Schema Document					***
101.CAL	Inline XBRL Taxonomy Extension Calculation Linkbase Document					***
101.DEF	Inline XBRL Extension Definition Linkbase Document					***
101.LAB	Inline XBRL Taxonomy Label Linkbase Document					***
101.PRE	Inline XBRL Taxonomy Extension Presentation Linkbase Document					***
104	Cover page Interactive Data File (formatted in Inline XBRL and contained in Exhibit 101) - The cover page interactive data file does not appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL document					***

† Portions of the exhibit(s) have been redacted in compliance with Regulation S-K Item 601(b)(10)(iv)
* Filed herewith
** Furnished herewith
*** Submitted electronically herewith

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned thereunto duly authorized.

KINIKSA PHARMACEUTICALS, LTD.

Date: May 6, 2021

By: /s/ Sanj K. Patel
Sanj K. Patel
Chief Executive Officer and Chairman of the Board of Directors

COMMERCIAL SUPPLY AGREEMENT

This Commercial Supply Agreement (including Exhibit 1 hereto, this "**Agreement**") is entered into as of February 26, 2021 (the "**Effective Date**") by and between:

Kiniksa Pharmaceuticals (UK), Ltd., a company incorporated under the laws of England and Wales having its registered office at Third Floor, 23 Old Bond Street, London W1S 4PZ, England ("**KINIKSA**"),

and

Regeneron Pharmaceuticals, Inc., a company organized under the laws of New York and having a principal place of business at 777 Old Saw Mill River Road, Tarrytown, New York 10591 (together with its Affiliates "**REGENERON**").

With each of KINIKSA and REGENERON collectively referred to as the "**Parties**" and individually as a "**Party**".

RECITALS

Whereas, REGENERON is a biopharmaceutical company engaged in the field of discovery, development, manufacture and commercialization of biopharmaceutical products;

Whereas, KINIKSA is engaged in the development, manufacture and commercialization of biopharmaceutical products;

Whereas, Kiniksa Pharmaceuticals, Ltd. and REGENERON have entered into a License Agreement dated as of September 25, 2017 (the "**License Agreement**");

Whereas, as of January 7, 2021, Kiniksa Pharmaceuticals, Ltd. has assigned the License Agreement to KINIKSA as part of a sale of assets relating to the Product;

Whereas, pursuant to the terms of the License Agreement, REGENERON intends to Manufacture and supply and KINIKSA intends to purchase Formulated Bulk Product and Filled Drug Product as set forth in this Agreement;

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.

ARTICLE 1
DEFINITIONS AND INTERPRETATION

1.1. Definitions

All capitalized terms not otherwise defined herein have the meanings ascribed to them set forth in the License Agreement. For the purposes of this Agreement, the following words and phrases shall have the

meanings ascribed to them below, even if different than the meanings ascribed to them in the License Agreement.

"Batch" shall mean a specific quantity of Formulated Bulk Product or Filled Drug Product that is intended to be of uniform character and quality, within specified limits, and is produced during the same cycle of manufacture, as defined by the applicable Batch Record and regulatory submissions. For purposes of this Agreement, (a) a Batch of Formulated Bulk Product is [***] Liter bioreactor production run and (b) a Batch of Filled Drug Product results in any number of vials within the validated batch size range (i.e., a minimum of [***] vials and a maximum of [***] vials, as of the Effective Date).

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

"**Binding Forecast"** has the meaning set forth in Section 5.3(a) below.

"**Clinical Supply Agreement"** shall mean the Clinical Supply Agreement between REGENERON and Kiniksa Pharmaceuticals, Ltd. effective September 27, 2017.

"CMO" or **"Contract Manufacturing Organization"** shall mean a company engaged by REGENERON to perform the filling of the Formulated Bulk Product.

"Delivery" shall mean the date REGENERON makes Product available to KINIKSA and the following have been achieved: (i) the Product has beforehand been released by REGENERON's quality control unit; (ii) KINIKSA has received the Batch Documentation (as defined in the Quality Agreement) signed by an authorized representative of REGENERON in accordance with the Quality Agreement, in addition to any other documentation required or specified in the Quality Agreement; and (iii) the representations and warranties set forth in Section 10.3 of this Agreement are true with respect to the Product being made available to KINIKSA hereunder.

"Delivery Date" shall mean the calendar day of delivery specified by KINIKSA in a Purchase Order in accordance with Section 5.4 and as agreed to by REGENERON.

"DS Facility" shall mean REGENERON's drug substance manufacturing site as identified in regulatory submissions for Product, which is, as of the Effective Date, located at 81 Columbia Turnpike Rensselaer, NY 12144.

"Filled Drug Product" shall mean Product filled in vials, before their labeling and packing for sale to the market in the Territory.

"Formulated Bulk Product" shall mean Product formulated into solution at the DS Facility that is ready for storage or shipment to KINIKSA or to KINIKSA's designee to allow for further processing.

"Force Majeure Event" shall have the meaning set forth in Section 12.6.

"Good Practices" shall mean compliance with the applicable standards contained in then-current "Good Laboratory Practices," "Good Manufacturing Practices" or "Good Clinical Practices," as promulgated by the FDA and all analogous guidelines promulgated by the EMA or the ICH, or other country Regulatory Authorities, as applicable.

"Law" or **"Laws"** shall mean any and all federal, state, local, national, and supra-national laws, statutes, rules, regulations, treaties, orders, judgments, injunctions or ordinances 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 the activities under this Agreement.

"Manufacture" or "**Manufacturing"** shall mean all activities directed to producing, manufacturing, processing, filling, finishing, quality assurance, quality control, testing and release, shipping and storage of Formulated Bulk Product or Filled Drug Product.

"**Manufacturing Process"** shall mean all the Manufacturing operations performed under the responsibility of REGENERON.

"Minimum Shelf Life" unless the Parties have agreed otherwise in writing, shall mean (a) Formulated Bulk Product with a minimum of [***] months remaining shelf life from the date made available to KINIKSA for Delivery, and (b) Filled Drug Product with a minimum of [***] months remaining shelf life from the date made available to KINIKSA for Delivery; *provided, however*, that in either case, if the total potential shelf life for Product is extended by an applicable Regulatory Authority, then the Parties shall discuss and consider in good faith extending the Minimum Shelf Life for such Product by an agreed upon amount and in accordance with the Quality Agreement and shall document any resulting agreement through a written amendment to this Agreement.

"Non-Conforming Product" shall mean, with respect to any Product: (a) any failure to conform to the Specifications set forth in the Quality Agreement; or (b) that the Product was not manufactured in accordance with GMP and applicable Law.

"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) [***]. Product shall either be Filled Drug Product or Formulated Bulk Product, unless otherwise agreed by the Parties. As used herein, Product shall also include any reference standard purchased by KINIKSA pursuant to Section 6.5.

"Purchase Order" shall mean a written KINIKSA purchase order which, upon acceptance in writing by REGENERON, shall constitute a non-cancellable, legally binding commitment on the part of KINIKSA to purchase the quantity of Product set forth in such purchase order, the form of which is attached hereto as Exhibit 1.

"Quality Agreement" shall mean the quality agreement entered into between the Parties, as may be amended from time to time, which (i) sets forth the Specifications, and the quality requirements for the Manufacture and supply of Product by REGENERON or on behalf of REGENERON to KINIKSA in accordance with Good Practices, and (ii) defines the roles and responsibilities of the Parties and their respective Affiliates and subcontractors with regard to importation in the European Union, quality testing, certification and other quality matters.

"Specifications" shall mean the mutually agreed and written list of tests, references to analytical procedures, and appropriate acceptance criteria or other criteria for tests according to which Product Manufactured by or on behalf of REGENERON hereunder must conform to be considered acceptable for its intended use.

"Term" shall have the meaning set forth in Section 9.1.

"Territory" means all the countries and territories of the world, other than the Retained Territory.

1.2. Certain Rules of Interpretation in this Agreement and the Exhibits

(a) The descriptive headings of Articles and Sections are inserted solely for convenience of reference and are not intended as complete or accurate descriptions of the content of such Articles or Sections.

(b) The use of words in the singular or plural, or with a particular gender, shall not limit the scope or exclude the application of any provision of this Agreement to such person or persons or circumstances as the context otherwise permits.

(c) Whenever a provision of this Agreement requires an approval or consent by a Party to this Agreement and notification of such approval or consent is not delivered within the applicable time limit, then, unless otherwise specified, the Party whose approval or consent is required shall be conclusively deemed to have withheld its approval or consent.

(d) In the event of any conflict between this Agreement and the Exhibits hereto, this Agreement shall prevail; *provided, however*, that if there is a conflict between this Agreement and the Quality Agreement on a quality-related matter, the Quality Agreement shall prevail. In the event of any conflict between this Agreement and the Exhibits hereto on the one hand, and the License Agreement on the other, the License Agreement shall control, unless there is explicit reference to the contrary herein, or if the conflict relates to quality matter, in which case the Quality Agreement shall control.

ARTICLE 2
PRODUCT COVERED BY THIS AGREEMENT

The purpose of this Agreement is to set forth the terms and conditions whereby, during the Term, KINIKSA shall order, and REGENERON shall use Commercially Reasonable Efforts to Manufacture (or have Manufactured) and supply (or have supplied), KINIKSA's requirements (as set forth in the Binding Forecast(s)) for Formulated Bulk Product or Filled Drug Product, as the case may be. KINIKSA may use Product purchased pursuant to this Agreement for Development or Commercialization activities. This Agreement applies to Purchase Orders placed pursuant to this Agreement after the Effective Date and does not apply to Purchase Orders placed pursuant to the Clinical Supply Agreement.

ARTICLE 3
GENERAL OBLIGATIONS

3.1 Manufacturing Requirements; Duty to Advise

(a) During the Term and subject to Section 8.14.1 of the License Agreement (Discontinuation of Formulated Product Manufacturing), REGENERON shall use Commercially Reasonable Efforts to (i) Manufacture the Formulated Bulk Product at the DS Facility, and (ii) as applicable, have the Formulated Bulk Product or Filled Drug Product Manufactured and delivered to KINIKSA, in each instance in accordance with (A) this Agreement, (B) the Quality Agreement (including the Specifications), (C) Good Practices, (D) the Purchase Order, and (E) any and all applicable Laws; and KINIKSA shall order, purchase and pay for such Formulated Bulk Product or Filled Drug Product manufactured in accordance with the terms of this Agreement and each respective Purchase Order.

(b) REGENERON shall promptly provide written notice to KINIKSA of any of the following events or occurrences, or any facts or circumstances reasonably likely to give rise to any of the following events or occurrences: (i) any failure by REGENERON to perform any of its material obligations under this Agreement; (ii) any delay in Delivery of Product; (iii) any defects or quality problems relating to Product; (iv) any Change of Control of REGENERON; (v) any deficiency or material non-conformity in the starting materials; or (vi) any failure by REGENERON, or its approved subcontractors providing Manufacturing services, to comply with applicable Law. In addition, REGENERON shall promptly notify

KINIKSA in writing of any change in REGENERON's authorized representatives that adversely affects any Product, Manufacturing, this Agreement or the Quality Agreement.

3.2 Product; Changes in Manufacturing

(a) All Product ordered by KINIKSA hereunder shall be Manufactured using materials provided or purchased by REGENERON.

(b) Except as otherwise specifically agreed in writing between the Parties, REGENERON shall be responsible for purchasing or having purchased or providing all raw materials for the Product to be Manufactured at [***] cost.

(c) Any change in the Manufacturing Process of the Product must be effected in accordance with the change control procedures as described in Section 8.8 (Product Changes) of the License Agreement and as provided for in the Quality Agreement.

(d) The Formulated Bulk Product will be Manufactured [***]. Any proposed change to [***] will [***] in accordance with Section 8.1.4 of the License Agreement (Decisions Regarding Manufacturing Transfer) and the Quality Agreement, [***].

(e) The Master Batch Record for the Manufacture of Product shall be prepared in accordance with the Quality Agreement.

3.3. Performance by Affiliates

Any of a Party's obligations identified herein or in the Quality Agreement can, at such Party's discretion, be performed by any of its Affiliates, and any of a Party's rights hereunder or under the Quality Agreement can also be executed by any of such Party's Affiliates, provided that, as contemplated by and without limitation of Sections 17.9.2, 17.9.3 and 17.9.4 of the License Agreement, such Affiliate agrees in writing to be bound by the terms and conditions of this Agreement.

ARTICLE 4
LICENCES / APPROVALS

Upon the terms and subject to the conditions hereof, each Party will use Commercially Reasonable Efforts to (a) obtain from the requisite Governmental Authorities any consents, licenses, permits, waivers, approvals, authorizations or orders required to be obtained or made by such Party in connection with the authorization, execution and delivery by such Party of this Agreement and the performance by such Party of its obligations hereunder, and (b) make all necessary filings, and thereafter make any other advisable submissions, required to be made by such Party under applicable Laws with respect to this Agreement or such Party's performance of its obligations hereunder. The Parties will reasonably cooperate with each other in connection with the making of all such filings, including by providing copies of all such non-confidential documents to the other Party prior to the filing and, if requested, by considering all reasonable additions, deletions or changes suggested by the other Party in connection therewith.

ARTICLE 5
FORECASTS AND DELIVERY

5.1 Supply Chain Management

Each Party shall, forthwith upon execution of this Agreement, appoint one (1) of its employees to be a point of contact responsible for acting as liaison between the Parties with regard to the supply chain

matters described in this Agreement. The points of contact shall meet (either in person or by phone) on a monthly basis or such other frequency as the points of contact may agree upon, but in any event not less frequently than every three (3) months, to review the current status of the supply relationship. Additional subject matter experts shall be available for participation in these meetings as needed. The points of contact will use Commercially Reasonable Efforts to address forecasting and production planning matters jointly.

5.2 Forecasts and Order Quantity

(a) In the [***] of each calendar quarter, KINIKSA will prepare and deliver to REGENERON for its review (in accordance with Section 5.3, below) a rolling long-term forecast for the estimated quantity of Product KINIKSA will require (i.e., specifying either Filled Drug Product and/or Formulated Bulk Product) for a minimum period of [***] months commencing on the month in which the forecast is delivered ("**Long-Term Forecast**"). Notwithstanding the foregoing, KINIKSA may choose to deliver the Long-Term Forecast more frequently than quarterly.

(b) The Parties acknowledge that as of the Effective Date, REGENERON's minimum campaign size of Formulated Bulk Product is [***] Batches. Given this minimum campaign size, KINIKSA is obligated to purchase at least [***] Batches for every manufacturing campaign that REGENERON runs for Formulated Bulk Product based on KINIKSA's Binding Forecast.

(c) Any Binding Forecast and Purchase Order that includes Formulated Bulk Product or Filled Drug Product must be in full Batch quantities, subject to the following:

(i) Subject to Section 5.2(b), the requirement to purchase in full Batch quantities means purchases in increments of one full Batch (e.g., KINIKSA may purchase one (1), or two (2), or three (3), etc. full Batches).

(ii) KINIKSA may achieve a full Batch quantity order of Formulated Bulk Product by placing a Purchase Order for a combination of Formulated Bulk Product and Filled Drug Product with the intent that the Filled Drug Product is converted from the same Batch of Formulated Bulk Product that is ordered in such Purchase Order.

(iii) KINIKSA shall take Delivery of a full Batch of Product on a single Delivery Date. Notwithstanding the foregoing, if KINIKSA places a Purchase Order for a combination of Formulated Bulk Product and Filled Drug Product as described in subsection (ii) above, then KINIKSA shall take Delivery of all of the Filled Drug Product on a single Delivery Date and KINIKSA shall take Delivery of all of the Formulated Bulk Product that remains on a single Delivery Date, which may be a different date than the date on which KINIKSA took delivery of the Filled Drug Product. KINIKSA may take Delivery of Filled Drug Product and Formulated Bulk Product on different Delivery Dates, even if KINIKSA ordered Filled Drug Product and Formulated Bulk Product in the same Purchase Order.

(iv) The Parties agree and acknowledge that actual yields of Product during Manufacturing may vary. In the instance where the actual yield of Product varies from the quantity specified in KINIKSA's Purchase Order, then KINIKSA will revise its Purchase Order (either upwards or downwards, as applicable) to reflect the actual quantity Manufactured by REGENERON.

(v) REGENERON shall not supply Product to any Third Party from a Batch that was Manufactured for KINIKSA pursuant to a Purchase Order.

(vi) For the sake of clarity, the requirements of this Section 5.2(c) do not apply to purchase orders for Product that KINIKSA placed pursuant to the Clinical Supply Agreement prior to the Effective Date.

(d) For the sake of clarity, except as described above in Section 5.2(c)(iv), REGENERON may not supply to KINIKSA quantities of Product in excess of KINIKSA's Purchase Order, unless otherwise authorized in writing by KINIKSA.

5.3 REGENERON's Forecast Obligations

(a) No later than [***] calendar days after receipt of KINIKSA's Long-Term Forecast, REGENERON shall accept or reject in writing such Long-Term Forecast. Upon REGENERON's written acceptance of a Long-Term Forecast, it shall become binding (the "**Binding Forecast**") with respect to (i) the Formulated Bulk Product, for a period of the first [***] months of such Long-Term Forecast, and (ii) the Filled Drug Product for a period of the first [***] months of such Long-Term Forecast.

(b) Notwithstanding Section 5.3(a), REGENERON shall have [***] Business Days to accept or reject the amounts set forth in the newly added (i.e., the last) time periods of each Binding Forecast, and REGENERON will [***] such amounts if they are consistent with the amounts set forth in the corresponding time periods of the Long-Term Forecast that was submitted and accepted by REGENERON in the prior calendar quarter.

(c) If REGENERON rejects any Long-Term Forecast, including any rejection relating to the Binding Forecast, then when REGENERON provides its written notice of rejection, it shall also provide a written explanation as to what was not acceptable. The Parties shall use Commercially Reasonable Efforts to reach a mutually agreeable solution, such as where possible, bringing forward or postponing a portion of the non-binding Long-Term Forecast, such that KINIKSA may submit a Long-Term Forecast that REGENERON will accept. For the sake of clarity, REGENERON may not reject any portion of the Binding Forecast that REGENERON had already accepted as part of a previously submitted and accepted Long-Term Forecast.

5.4 Purchase Orders

(a) KINIKSA shall be required to issue a Purchase Order quarterly for Product in a quantity consistent with the current Binding Forecast within [***] calendar days of REGENERON'S acceptance of said Binding Forecast; *provided, however*, that, except as described in Section 5.2(c)(iv), any Purchase Order shall not adjust the quantity or change other specified details for any months covered by a Purchase Order that has already been issued by KINIKSA and accepted by REGENERON.

(b) REGENERON shall not reject any Purchase Order submitted by KINIKSA that is consistent in quantity and Delivery Date with a Binding Forecast REGENERON has previously accepted. REGENERON shall accept or reject each Purchase Order within [***] Business Days after REGENERON's receipt of such Purchase Order.

(c) Each Purchase Order shall detail the Purchase Order number, the REGENERON Item Master Number, the Product name, Product quantity, and the Delivery Date.

(d) In the event that KINIKSA issues a Purchase Order for Product that is less than what was set forth in an accepted Binding Forecast, [***], KINIKSA shall [***]; *provided, however* that such obligation to [***] is subject to REGENERON's obligation to fill the excess capacity or use other mitigating measures pursuant to Section 7.5 below. Notwithstanding the foregoing, [***], KINIKSA may elect to take delivery of the Product at a mutually acceptable date that conforms with Specifications in

accordance with the Binding Forecast and will be responsible for the corresponding [***] of such Product.

5.5 Delivery

(a) REGENERON shall use Commercially Reasonable Efforts to Manufacture and supply on the Delivery Date the quantity of Product specified in each Purchase Order. Notwithstanding the foregoing, for any given Purchase Order for Filled Drug Product, REGENERON may, in its sole discretion, with at least [***] days advanced written notice, deliver the Filled Drug Product up to [***] days earlier than the Delivery Date set forth in such Purchase Order. Except as provided in this Section 5.5(a), REGENERON shall not Deliver Product prior to the Delivery Date specified in a Purchase Order unless agreed upon by both Parties. REGENERON's obligation to supply on any Delivery Date shall be deemed satisfied if the Product is delivered within [***] calendar days thereof.

(b) If, despite using Commercially Reasonable Efforts to Manufacture and supply, any circumstances occur that are reasonably likely to result in an inability of REGENERON to meet the Delivery Date or quantity of Product specified in a Purchase Order which has been issued in accordance with Section 5.4 (including a Force Majeure Event or pursuant to Section 5.5(c) below), REGENERON shall as promptly as possible after becoming aware of such circumstance provide written notice to KINIKSA thereof, in which REGENERON proposes to KINIKSA a new delivery date and/or quantity. For any such failure in the timely delivery of ordered quantities, (i) REGENERON shall continue to use Commercially Reasonable Efforts to Manufacture and supply such quantity of Product as close as possible to KINIKSA's original Delivery Date specified in the applicable Purchase Order and (ii) KINIKSA shall have the option to cancel the corresponding Purchase Order (or portion thereof as the case may be), or to accept REGENERON's proposal of a new delivery date for such Purchase Order (or portion thereof as the case may be), which shall then be deemed to be the new Delivery Date with regard to such Purchase Order (or portion thereof). Nothing contained in this Section 5.5 limits KINIKSA's rights under Section 8.15 of the License Agreement in the event of a Manufacturing Technology Transfer Event (as defined in the License Agreement).

(c) Notwithstanding the foregoing or any other provision, in the event that REGENERON is not able to Manufacture or Deliver Product in accordance with the Binding Forecast or any Purchase Order [***], REGENERON shall notify KINIKSA of such disruption to the supply thereof, stating the period of time that the disruption is expected to continue. In such circumstances, the Parties will discuss in good faith a revised schedule of Manufacture and Delivery; provided that REGENERON shall not be held liable or responsible to KINIKSA nor be deemed to have defaulted under or breached this Agreement for such failure or any delay in Manufacturing or Delivery of such Product.

(d) Samples shall be delivered with each Batch, in accordance with the Quality Agreement.

5.6 Shipment and Title

(a) REGENERON shall arrange for Delivery to KINIKSA, or an Affiliate or designee of KINIKSA designated by written notice to REGENERON, Product Manufactured by REGENERON or its CMO as provided hereunder. Product shall be released for shipment by REGENERON and KINIKSA in accordance with this Agreement and the Quality Agreement. REGENERON shall further prepare and send all documentation required by applicable Laws, this Agreement and the Quality Agreement. Title, possession, risk of loss, risk of damage and all forward costs and expenses shall pass to and be borne by KINIKSA upon [***]. Prior to such [***], title, possession, risk of damage and loss shall remain with REGENERON and REGENERON shall insure all Product for full replacement value.

(b) Freight terms shall be [***] (Incoterms 2020) [***] for Formulated Bulk Product and [***] for Filled Drug Product, as applicable to Product ordered. REGENERON shall retain representative

samples of Product solely for record keeping, testing obligations or testing in the event of an alleged Batch non-conformity and regulatory purposes. KINIKSA shall provide for shipping of each Batch of Product on the Delivery Date, unless otherwise agreed between the Parties or as such Delivery Date may be adjusted pursuant to Section 5.5(a). In the event of any delay by KINIKSA in shipping or accepting of Product in accordance with this Section 5.6, the Parties shall work together in good faith to reschedule the Delivery Date, provided that [***] effective upon expiration of [***] calendar days after the originally scheduled Delivery Date.

ARTICLE 6
QUALITY CONTROL OF PRODUCTS

6.1 Personnel and Facilities

(a) REGENERON shall provide, and ensure that its CMOs provide, the necessary expertise, adequately skilled and trained personnel, facility and equipment to carry out its obligations under this Agreement, and it shall use Commercially Reasonable Efforts to maintain all Manufacturing facilities in compliance with Good Practices, applicable Law and the manufacturing license for Product.

(b) Throughout the Term, REGENERON will provide directly, or indirectly through written contract with a CMO, appropriately qualified and trained personnel, adequate premises and space, suitable equipment and services, correct materials, containers and labels, suitable storage, and the knowledge and experience to carry out its obligations under this Agreement.

(c) REGENERON owns or lawfully controls the DS Facility. During the Term, REGENERON will ensure that that the DS Facility and any CMO Manufacturing facility is maintained in accordance with Good Practices and all applicable Laws and in such condition as to enable Product to be Manufactured in compliance with Good Practices, all applicable Law, and the Quality Agreement.

(d) The terms and conditions of KINIKSA's audit rights and rights to inspect the facilities involved in Manufacturing the Formulated Bulk Product are provided in the Quality Agreement.

(e) During Manufacture of Formulated Bulk Product, REGENERON will permit [***] to observe GMP manufacturing of intermediates and Formulated Bulk Product. [***] must be accompanied by REGENERON personnel at all times and abide by all of REGENERON's policies and other requirements. In advance of [***] visit, KINIKSA must obtain written permission from REGENERON in order to be allowed [***] or to send [***] to REGENERON's site, provided that such permission shall not be unreasonably withheld or delayed.

(f) During Manufacture of Filled Drug Product, REGENERON shall take reasonable commercial efforts to ensure that REGENERON's subcontractor will permit a REGENERON "person in plant" to oversee GMP Manufacturing Activities at the subcontractor's facility. To the extent mutually agreeable with such subcontractor, REGENERON and KINIKSA, a KINIKSA employee may accompany the REGENERON "person in plant" for such portion or portion(s) of oversight, as applicable.

6.2 Work Output

All information, data, documentation, and reports produced by or on behalf of REGENERON in the conduct of Manufacturing Product or all other GMP documentation relating to Product will be prepared by REGENERON using REGENERON's SOPs.

6.3 Quality Control; Disputes

(a) REGENERON shall meet or exceed quality standards for the Product as set forth in the Quality Agreement. REGENERON shall perform quality inspections of Product before Delivery and shall certify inspection results in the manner agreed upon in the Quality Agreement.

(b) REGENERON shall operate the DS Facility in compliance with applicable Laws and any requirements provided for in the Quality Agreement for all aspects of Manufacturing (including but not limited to testing, holding, and delivery of the Product produced for KINIKSA). For clarity, this shall include the obligation for REGENERON to operate the DS Facility as compliant with (i) the U.S. Federal Food, Drug and Cosmetics Act, as amended (21 U.S.C. et seq.), (ii) U.S. regulations found at 21 CFR Parts 11, 210, 211, 600, & 610, (iii) ICH Q7 GMP Guidance for Active Pharmaceutical Ingredients, (iv) EC Directive 2003/94/EC, (v) the EC Guide to Good Manufacturing Practice for Medicinal Products and Division 2 of the Food and Drug Regulations (Canada), (vi) the latest Health Canada, FDA and EMA guidance documents all pertaining to manufacturing and quality control practice, and (vii) the good manufacturing practices required by any other Regulatory Authority to the extent such required practices do not contradict or negate the requirements or guidance documents issued by FDA or EMA, all as updated, amended and revised from time to time and applicable to the Product.

(c) KINIKSA shall inform REGENERON in writing of any visible damage or partial or full loss of Product during shipment or obvious deviation (including without limitation from the representations and warranties set forth in Section 10.3 hereof) or non-conformity based on visual inspection within [***] Business Days following receipt of such shipment at KINIKSA's designee. To the extent KINIKSA performs testing or has testing performed (as opposed to relying on REGENERON's Certificate of Analysis), KINIKSA shall inform REGENERON in writing of any non-conformity to the tested Specifications within [***] calendar days after Delivery. Product that is not rejected within the time periods set forth in this subsection will be deemed to have been accepted by KINIKSA; *provided, however*, that KINIKSA's acceptance will not be deemed to be a waiver or limitation of REGENERON's obligations pursuant to this Agreement or the License Agreement (or any breach thereof), including those obligations with respect to REGENERON's Warranties and Duty to Indemnify.

(d) If the Parties are unable to agree as to whether any Product is Non-Conforming Product, or to determine the cause of any deviation or 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 deviation or Non-Conforming Product. Such outside testing laboratory or consultant will determine, using samples of the applicable Batch of Product, whether the Batch of Product is deviating or 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 deviation or non-conformity. The fees and expenses of such testing shall be considered Other Shared Expenses.

(e) If, after Delivery of any Product supplied by REGENERON, either Party finds a previously undetected deviation or non-conformity or that such Product was adulterated or misbranded, the discovering Party shall promptly notify the other Party.

(f) Notwithstanding anything to the contrary contained herein, KINIKSA shall return any non-conforming shipment of Product to REGENERON or otherwise dispose thereof, as REGENERON shall direct. REGENERON shall use Commercially Reasonable Efforts to Manufacture, have Manufactured and deliver to KINIKSA, as promptly as possible, a quantity of Product replacing such Non-Conforming Product.

(g) Without limiting KINIKSA's rights contained in Section 14.2 of the License Agreement (Indemnity by Regeneron) or Section 5.5(a) of this Agreement, and except for REGENERON's liability for recalls and withdrawals as set forth in Section 7.8 of the License Agreement (Recalls and Other Corrective

Actions), KINIKSA's [***], and REGENERON's (or its Affiliate's) [***] with respect to defective or Non-Conforming Products or failure to supply Product in accordance with the terms of the Quality Agreement or this Agreement shall be the [***]. For clarity and, if applicable, KINIKSA shall also have the right to effectuate a Manufacturing Technology Transfer in the event of a Manufacturing Technology Transfer Event consistent with the terms set forth in the License Agreement.

(h) In the event of Batch failure, refer to Section 8.11 of the License Agreement (Defective Product).

6.4 Limitation Period for Certain Claims

The limitation period for all claims by KINIKSA related to defective or Non-Conforming Products is [***] months from the Delivery Date, provided that this Section 6.4 shall not alter the time periods set forth in Section 6.3 within which KINIKSA must notify REGENERON of Product defects or non-conformity or the effect of a failure by KINIKSA to timely give such notice.

6.5 Reference Standards

From time to time at KINIKSA's reasonable request, REGENERON shall provide to KINIKSA the list of suppliers for reference standards required for the testing by KINIKSA of the Product, and, in case there is no commercially available supplier of any given reference standard, REGENERON shall supply KINIKSA with such reasonable quantities of reference standard as reasonably requested by KINIKSA, at a price equal to REGENERON's Fully-Burdened Costs and under reasonable lead-time as agreed between the Parties.

ARTICLE 7
PRICE AND PAYMENT TERMS

7.1 Price

The price for the specific Product delivered by REGENERON to KINIKSA pursuant to a Purchase Order that meets all of the requirements of this Agreement shall be the [***] for such Product and paid in accordance with the [***], as set forth in Section 9.4.2(a) of the License Agreement (Kiniksa Payment to Regeneron for Product Supplied under the Supply Agreement) and the [***] as set forth in Section 9.4.2(b) of the License Agreement [***] (if any) for such period, [***], subject however, to any adjustments set forth in Section 6.3 hereof.

7.2 Invoicing

In accordance with Section 8.6 of the License Agreement (Price for Product Supplied by Regeneron), for Product ordered pursuant to a Purchase Order submitted in accordance with Section 5.4 (Purchase Orders), REGENERON shall invoice KINIKSA for the [***] concurrently with the Delivery of the Product to Kiniksa.

7.3 Payment Terms

Payment terms applicable to Product deliveries and invoices shall be determined as set forth in Section 9.4.2 of the License Agreement (Reimbursement of Manufacturing Costs). REGENERON shall issue invoices for Product upon Delivery of such Product. Notwithstanding the foregoing, until the [***] year anniversary of the Effective Date, if REGENERON Delivers [***] between [***] and [***] days prior to the Delivery Date set forth in the Purchase Order pursuant to Section 5.5(a), then KINIKSA shall pay for such [***] no later than [***] days after KINIKSA's receipt of an invoice for such [***].

7.4 Late Payments

Any late payments for Products delivered or produced hereunder shall be subject to Section 9.7 of the License Agreement (Late Payments) and Section 9.9 of the License Agreement (Resolution of Payment Disputes).

7.5 Cancellation of Purchase Orders

The Purchase Order acceptance by REGENERON of any quantity of Product ordered shall constitute a legally binding commitment on the part of KINIKSA to purchase the quantity of Product set forth therein; *provided, however*, that in the event that KINIKSA cancels all or any portion of any Purchase Orders, KINIKSA may be obligated to pay to REGENERON as set forth above in Section 5.4(d). REGENERON will, however, in good faith, exert Commercially Reasonable Efforts to fill the excess capacity or use other mitigating measures in order to reduce the Manufacturing Costs for which KINIKSA might be liable.

7.6 Currency

All payments under this Agreement shall be made in US Dollars.

7.7 Taxes

Taxes shall be addressed in accordance with the License Agreement.

ARTICLE 8
CONFIDENTIALITY

Confidentiality shall be governed under the terms of the License Agreement in the confidentiality provisions and the related definitions shall be deemed to also refer to this Agreement for purposes of this ARTICLE 8.

ARTICLE 9
TERM AND TERMINATION

9.1 Term

This Agreement shall commence on the Effective Date and shall remain in force for the earlier to occur of (i)expiration or termination of the License Agreement or (ii) the date of the completion of a Manufacturing Technology Transfer Event as set forth in a Technology Transfer Agreement, which may take the form of a Statement of Work, attachment to this Agreement, or other separate agreement.

9.2 Termination

Except as otherwise provided in Article 16 of the License Agreement (Term and Termination) and Section 12.11 of this Agreement (Survival), 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.

ARTICLE 10
SUPERIORITY; WARRANTIES; INDEMNITY

10.1 Superiority

No provision in KINIKSA's Purchase Order, or in REGENERON's confirmation or acceptance of a Purchase Order, general conditions of sale or invoice, which may purport to impose different conditions upon KINIKSA or REGENERON, shall modify or otherwise alter the terms of this Agreement.

10.2 Warranties and Limitations of Liability

In addition to the additional warranties and any limitation of liability terms included herein, the warranties and limitations of liability of REGENERON and KINIKSA shall be governed by the warranty and limitation of liability terms set forth in the License Agreement.

10.3 General Warranties

REGENERON represents and warrants that (a) Product supplied under this Agreement to KINIKSA (i) shall be Manufactured in accordance with the Specifications and quality requirements set forth in the Quality Agreement and all Laws, including GMP, (ii) shall be free of any defects in any materials or workmanship, (iii) shall be stored and supplied in conformity with the Specifications and all Laws, including GMP, (iv) shall meet the Minimum Shelf Life requirements for such Product at the time of Delivery, provided that if REGENERON Delivers Filled Drug Product early pursuant to Section 5.5(a), such Filled Drug Product must meet the Minimum Shelf Life requirement as of the Delivery Date set forth in the Purchase Order, and (v) 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 Laws, including GMP; (b) it will not introduce any materials not provided in the Specifications that would cause the applicable Product to be adulterated or misbranded within the meaning of Sections 501 or 502 of the U.S. Federal Food, Drug and Cosmetics Act and the regulations promulgated thereunder; and (c) it shall perform all obligations hereunder in compliance with all Laws and industry standards of workmanship and professionalism.

10.4 Indemnity & Insurance

Indemnity and insurance shall be governed by the terms set forth in Article 14 of the License Agreement (Indemnity).

ARTICLE 11
ASSIGNMENT

Subject to Section 3.3 (Performance by Affiliates) hereof, neither Party shall transfer, assign or delegate this Agreement, or any rights or obligations hereunder, in whole or in part, without the other Party's prior written consent, except (a) to an Affiliate of the transferring, assigning or delegating Party, or (b) in connection with the assignment of the License Agreement in compliance with Section 17.9.1 of the License Agreement (Assignment to Third Parties by Kiniksa). Any purported transfer, assignment or delegation not in compliance with this Article 11 shall be void and of no effect.

ARTICLE 12
MISCELLANEOUS

12.1 Severability

If any provision of this Agreement or the application thereof to any Party or circumstances is held invalid, such invalidity shall not affect other provisions or applications of this Agreement which can be given effect without the invalid provision or application and to this end the provisions or applications of this Agreement are declared to be severable. In lieu of any invalid, prohibited or unenforceable provision or

application thereof, the Parties or a court (if applicable) shall substitute suitable or equitable terms to carry out the intent of this Agreement.

12.2 Amendments; Waivers

Any waiver, amendment or modification of this Agreement is unenforceable unless made in a written document executed by duly authorized representatives of both KINIKSA and REGENERON. 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.

12.3 Relationship of the Parties

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.

12.4 Parties in Interest

This Agreement shall inure to the benefit of and be binding upon the Parties and their respective permitted successors and assigns. Nothing in this Agreement, express or implied, is intended to confer on any person other than the Parties, or their respective successors or assigns, any rights, remedies, obligations or liabilities under or by reason of this Agreement.

12.5 Compliance with Laws.

In the performance of this Agreement, both Parties agree to comply with all applicable Laws.

12.6 Force Majeure

Without limiting the terms of Section 8.13 (Notification and Discussion of Supply Issues) and Section 8.14 (Manufacturing Technology Transfer Event) of the License Agreement, a delay or failure of either Party to perform its obligations under this Agreement will be excused to the extent that the delay or failure was caused directly by an event beyond such Party's control, without such Party's fault or negligence and that by its nature could not have been foreseen by such Party or, if it could have been foreseen, was unavoidable (which events may include natural disasters, embargoes, explosions, riots, wars or acts of terrorism, pandemic (including without limitation the COVID-19 pandemic and any iterations of such pandemic) (each, a "**Force Majeure Event**"). A Party's financial inability to perform, changes in cost or availability of materials, components or services, market conditions or supplier actions or contract disputes will not excuse performance under this Section 12.6. REGENERON shall give KINIKSA prompt written notice of any event or circumstance that is reasonably likely to result in a Force Majeure Event and the anticipated duration of such Force Majeure Event. REGENERON shall use

Commercially Reasonable Efforts to end the Force Majeure Event, ensure that the effects of any Force Majeure Event are minimized and resume full performance under this Agreement.

12.7 Notices

Any consent, notice or report required or permitted to be given or made under this Agreement by one of the Parties to the other shall be in writing, delivered personally or by first class air mail or nationally recognized courier, postage prepaid (where applicable), addressed to such other Party at its address indicated below, or to such other address as the addressee shall have last furnished in writing to the addressor and (except as otherwise provided in this Agreement) shall be effective upon receipt by the addressee.

If to REGENERON:

Regeneron Pharmaceuticals, Inc.
777 Old Saw Mill River Road
Tarrytown, New York 10591
Attention: General Counsel

With a copy to:

Regeneron Pharmaceuticals, Inc.
1 Global View
Troy, NY 12180
Attention: Assistant General Counsel

If to KINIKSA:

Kiniksa Pharmaceuticals (UK), Ltd.
Third Floor
23 Old Bond Street
London W1S 4PZ
England
Attn: Legal Department

With a copy to:

Kiniksa Pharmaceuticals Corp.
100 Hayden Avenue
Lexington, MA 02421
Attn: Legal Department

12.8 Governing Law and 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 of the License Agreement (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.

12.9 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 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.

12.10 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.

12.11 Survival.

Subject to the limitations and other provisions of this Agreement: (a) the representations and warranties of the Parties contained herein will survive the expiration or earlier termination of this Agreement; and (b) Article 1 (Definitions and Interpretation), Article 8 (Confidentiality), Article 9 (Term and Termination), Article 10 (Superiority; Warranties; Indemnity), Article 12 (Miscellaneous), Section 6.3(g), and Section 6.4 (Limitation Period for Certain Claims) of this Agreement, as well as any other provision that, in order to give proper effect to its intent, should survive such expiration or termination, will survive the expiration or earlier termination of this Agreement. All other provisions of this Agreement will not survive the expiration or earlier termination of this Agreement.

12.11 Equitable 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.

12.12 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.

[Remainder of page intentionally left blank; Signature page follows]

IN WITNESS WHEREOF, the Parties have caused this Agreement to be signed by their duly authorized corporate officers, on the dates indicated below.

Kiniksa Pharmaceuticals (UK), Ltd.	**Regeneron Pharmaceuticals, Inc.**
__/s/ Ross Moat_____	__/s/ Scott Oberman_____
Name: Ross Moat	Name: Scott Oberman
Title: Director	Title: Vice President, Supply Chain Operations

Exhibit 1: Purchase Order

Order To:			Ship to:			Bill To:		
PO #		PO Date	Payment Terms			Shipping Terms		
Special Instructions:								
Country	Item #	Description	Qty.	Unit Size	Unit Price	Total	Notes	Delivery Date
		COMMERCIAL PURCHASE ORDER TOTAL				$0		

AMENDMENT NO. 1

THIS AMENDMENT NO. 1 ("**Amendment No. 1**") to that certain License Agreement dated as of September 25, 2017 (the "**Original Agreement**") by and between Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company and having an address at Clarendon House, 2 Church Street, Hamilton HM 11, Bermuda ("**Kiniksa**"), and 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**") is effective as of October 29, 2020. Each of Regeneron and Kiniksa shall be referred to herein individually as a "**Party**" and collectively as the "**Parties**". Unless otherwise defined herein, all capitalized terms shall have the meanings ascribed to them in the Original Agreement.

WHEREAS, Kiniksa and Regeneron desire to amend the Original Agreement to correct errors and reflect the true intent of the Parties;

NOW, THEREFORE, in consideration of the mutual undertakings herein contained, the Parties hereto agree as follows:

1. Amendment. The Original Agreement is hereby amended as follows:

    a. In Section 9.5.1, the reference to Section 1.189(f) of the definition of Shared Commercial Expenses shall be replaced with a reference to Section 1.189(g) of the definition of Shared Commercial Expenses so that the language reads:

    "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(g) of the definition of Shared Commercial Expenses for any Contract Year:"

    b. In Section 9.5.3(a), the reference to [***]

    "If the JSC approves a Subsequent Indication [***] so approved."

2. No Other Amendments. Except to the extent amended hereby, all of the terms, provisions and conditions set forth in the Original Agreement are hereby ratified and confirmed and shall remain in full force and effect. The Original Agreement and this Amendment No. 1 shall be read and construed together as a single agreement and the term "Agreement" as used shall henceforth be deemed a reference to the Original Agreement as amended by this Amendment No. 1.

3. Counterparts. This Amendment No. 1 may be signed in any number of counterparts, each of which shall be deemed to be an original and all of which together shall constitute one and the same instrument.

<Signature Page to Follow>

[***] Certain information in this document has been excluded pursuant to Regulation S-K, Item 601(b)(10). Such excluded information is not material and would likely cause competitive harm to the registrant if publicly disclosed.

IN WITNESS WHEREOF, duly authorized representatives of the Parties have executed this Amendment No. 1 as of the date first set forth above.

KINIKSA PHAMACEUTICALS, LTD.	**REGENERON PHARMACEUTICALS, INC.**
By: /s/ Thomas W. Beetham	**By:** /s/ Kerry Reinertsen
Name: Thomas W. Beetham	**Name:** Kerry Reinertsen
Title: Executive Vice President	**Title:** SVP, Strategic Alliances

[***] Certain information in this document has been excluded pursuant to Regulation S-K, Item 601(b)(10). Such excluded information is not material and would likely cause competitive harm to the registrant if publicly disclosed.

AMENDMENT NO. 2

THIS AMENDMENT NO. 2 ("**Amendment No. 2**") to that certain License Agreement dated as of September 25, 2017, and amended on October 29, 2020 (together referred to as the "**Agreement**") by and between Kiniksa Pharmaceuticals (UK), Ltd., a company incorporated under the laws of England and Wales having its registered office at Third Floor, 23 Old Bond Street, London W1S 4PZ, United Kingdom ("**Kiniksa**") and 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**") is effective as of the date of last signature below. Each of Regeneron and Kiniksa shall be referred to herein individually as a "**Party**" and collectively as the "**Parties**". Unless otherwise defined herein, all capitalized terms shall have the meanings ascribed to them in the Agreement.

WHEREAS, on January 7, 2021, Kiniksa Pharmaceuticals, Ltd. assigned the Agreement to its wholly-owned subsidiary Kiniksa Pharmaceuticals (UK), Ltd.;

WHEREAS, Kiniksa and Regeneron desire to amend the Agreement to add data protection langauge;

NOW, THEREFORE, in consideration of the mutual undertakings herein contained, the Parties hereto agree as follows:

4. Amendment. The Agreement is hereby amended as follows:

A new Section 12.6.4 is hereby added to the Agreement as follows:

"12.6.4 **Data Protection Law.** Each Party shall comply with Data Protection Law in performing its obligations under this Agreement. Any processing of personal data shall be for the Term of the Agreement, for the purpose of Developing or Commercializing Product, and include categories of personal data such as a Party's employee details and related information. When processing personal data on behalf of Regeneron, Kiniksa shall: (i) only process personal data on Regeneron's written instructions; (ii) ensure that all Kiniksa personnel who have access to such personal data are subject to suitable confidentiality obligations; (iii) implement and maintain technical and organizational measures to prevent a breach related to such personal data, and in the event of such a breach, Kiniksa shall notify Regeneron without undue delay and promptly undertake all remediation efforts necessary to rectify the personal data breach and prevent its recurrence; (iv) provide such assistance as Regeneron may reasonably require to meet its obligations under Data Protection Law (including the provision of information, responding to data subject and government requests and allowing for audits); (v) delete or return all personal data on Regeneron's written request; (vi) not subcontract such processing without Regeneron's prior written consent, which consent may not be unreasonably withheld or delayed, and Kiniksa shall remain fully liable for any of its subcontractors; and (vii) not transfer personal data from one jurisdiction to any other jurisdiction without Regeneron's prior written consent, which consent may not be unreasonably withheld or delayed. In this Section: (i) "**Data Protection Law**" means, all laws, rules and regulations, including any national implementing legislation relating to privacy and data protection; and (ii) "data subject", "personal data", "personal data breach" and "processing" will be construed in accordance with the respective Data Protection Law."

5. No Other Amendments. Except to the extent amended hereby, all of the terms, provisions and conditions set forth in the Agreement are hereby ratified and confirmed and shall remain in full force and effect. The Agreement and this Amendment No. 2 shall be read and construed together

as a single agreement and the term "Agreement" as used shall henceforth be deemed a reference to the Agreement as amended by this Amendment No. 2.

6. Counterparts. This Amendment No. 2 may be signed in any number of counterparts, each of which shall be deemed to be an original and all of which together shall constitute one and the same instrument.

IN WITNESS WHEREOF, duly authorized representatives of the Parties have executed this Amendment No. 2 as of the dates set forth below.

KINIKSA PHAMACEUTICALS (UK), LTD.	**REGENERON PHARMACEUTICALS, INC.**
By: /s/ Ross Moat	**By:** /s/ Kerry Reinertsen
Name: Ross Moat	**Name:** Kerry Reinertsen
Title: Director	**Title:** SVP, Strategic Alliances
Date: Apr 14, 2021	**Date:** Apr 13, 2021

[***] Certain information in this document has been excluded pursuant to Regulation S-K, Item 601(b)(10). Such excluded information is not material and would likely cause competitive harm to the registrant if publicly disclosed.

EMPLOYMENT AGREEMENT

This Employment Agreement (this "Agreement") is effective as of April 1, 2021 (the "Effective Date"), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the "Company"), and Mark Ragosa (the "Employee").

WHEREAS, the operations of the Company and its Affiliates (as defined below) are a complex matter requiring direction and leadership in a variety of arenas;

WHEREAS, the Employee possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates; and

WHEREAS, wishes to employ the Employee on the terms and conditions set forth in this Agreement, and the Employee wishes to be employed under such terms and conditions.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual promises, terms, provisions and conditions set forth in this Agreement, the Company and Employee hereby agree:

1. Definitions. Words or phrases that are initially capitalized or are within quotation marks shall have the meanings provided in this Section and as provided elsewhere herein. For purposes of this Agreement, the following definitions apply:

(a) "Affiliates" shall mean all persons and entities directly or indirectly controlling, controlled by or under common control with the Company, where control may be by management authority, contract or equity interest.

(b) "Cause" shall mean:

(i) The Employee's gross negligence or willful misconduct in performance of Employee's duties to the Company, where such gross negligence or willful misconduct has resulted in or reasonably could result in material damage to the Company or any of its Affiliates or successors; or

(ii) The Employee's commission of any act of fraud, embezzlement or professional dishonesty with respect to the business of the Company or any of its Affiliates; or

(iii) The Employee's commission of a felony or crime involving moral turpitude; or

(iv) The Employee's material breach of any provision of this Agreement or any other written agreement between Employee and the Company; or

(v) The Employee's failure to comply with lawful directives of the Company, which has caused or which reasonably could cause damage to the Company or any of its Affiliates or successors.

(c) "Change in Control" shall mean:

(i) a sale of all or substantially all of the Parent's assets; or

(ii) any merger, consolidation or other business combination transaction of the Parent with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the shares of voting capital shares of the Parent outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital shares of the surviving entity) a majority of the total voting power represented by the shares of voting capital shares of the Parent (or the surviving entity) outstanding immediately after such transaction; or

(iii) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital shares of the Parent. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur:

(A) on account of the acquisition of shares of voting capital shares by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Parent's shares of voting capital shares in a transaction or series of related transactions that are primarily a private financing transaction for the Parent, or

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the "Subject Person"), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operation of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

(d) "Code" shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.

(e) "Employee Benefit Plan" shall have the meaning ascribed to such term in Section 3(3) of the Employee Retirement Income Security Act of 1974, as amended.

(f) "Confidential Information and Non-Competition Agreement" shall mean the Employee Proprietary Information, Inventions Assignment, Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of May 10, 2018.

(g) "Parent" shall mean the Company's parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(h) "Parent Board" shall mean the board of directors of the Parent.

(i) "Person" shall mean an individual, a corporation, a limited liability company, an association, a partnership, an estate, a trust and any other entity or organization, other than the Company or any of its Affiliates.

2. Acceptance and Term. Subject to the terms and conditions set forth in this Agreement, the Company hereby offers, and the Employee hereby accepts, employment and/or continuing employment on an at-will basis. Subject to earlier termination as hereinafter provided, the Employee's employment shall continue until terminated pursuant to Section 5 hereof (the "Term").

3. Position, Duties and Responsibilities.

(a) During the Term, the Employee shall initially serve the Company as its Senior Vice President, Chief Financial Officer and shall initially report to the Chief Executive Officer. During the Term, the Employee shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of Employee's position.

(b) During the Term, the Employee shall devote Employee's full business time and Employee's best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee's duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

(c) Immediately upon termination of Employee's employment with the Company for any reason, Employee will be deemed to resign any and all positions held by Employee, whether as an officer or director of the Company, the Parent or any Affiliate of the Company, or as a member of any committees thereof.

4. Compensation and Benefits. As compensation for all services performed by the Employee during the Term and subject to the Employee's performance of Employee's duties and obligations to the Company and its Affiliates, pursuant to this Agreement or otherwise, the Company shall provide the Employee with the following compensation and benefits:

(a) Base Salary. The Company shall pay the Employee an annual base salary of $423,000, payable in accordance with the Company's standard payroll practices and procedures and subject to change from time-to-time in the Company's sole discretion (such base salary, as from time-to-time changed, the "Base Salary").

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus ("Discretionary Annual Bonus") with an initial target level of 40% of Employee's Base Salary (the "Target Bonus"). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15th of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

(c) Vacation. During the Term, the Employee shall be entitled to earn vacation at the rate of four (4) weeks per year, to be taken at such times and intervals as shall be determined by the Employee, subject to the reasonable business needs of the Company. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

(d) Other Benefits. During the Term, the Employee shall be entitled to participate, to the extent eligible, in any and all Employee Benefit Plans from time-to-time in effect for employees of the Company generally, except to the extent any such Employee Benefit Plan is in a category of benefit otherwise provided to the Employee under this Agreement (e.g., a severance pay plan). Such participation shall be subject to the terms of the applicable plan documents and generally applicable Company policies. The Company may alter, modify, add to or discontinue its Employee Benefit Plans at any time as it, in its sole judgment, determines to be appropriate, without recourse by the Employee.

(e) Business Expenses. The Company shall pay or reimburse the Employee for all reasonable business expenses incurred or paid by the Employee in the performance of Employee's duties and responsibilities hereunder, subject to reasonable substantiation and documentation and the Company's standard expense reimbursement policies and procedures.

5. Termination of Employment and Severance Benefits. The Employee's employment with the Company shall terminate under the following circumstances:

(a) Death. In the event of the Employee's death, the Employee's employment hereunder shall immediately and automatically terminate.

(b) Disability.

(i) The Company may terminate the Employee's employment hereunder, upon notice to the Employee, in the event that the Employee becomes disabled during Employee's employment hereunder through any illness, injury, accident or condition of either a physical or psychological nature and, as a result, is unable to perform substantially all of Employee's duties and responsibilities hereunder, notwithstanding the provision of any reasonable accommodation, for ninety (90) consecutive days.

(ii) The Parent Board may designate another employee to act in the Employee's place during any period of the Employee's disability. Notwithstanding any such designation, the Employee shall continue to receive the Base Salary in accordance with Section 4(a) and benefits in accordance with Section 4(d), to the extent permitted by the then-current terms of the applicable benefit plans, until the Employee becomes eligible for disability income benefits under any disability income plan or until the termination of Employee's employment, whichever shall first occur.

(iii) While receiving disability income payments under any disability income plan, the Employee shall not be entitled to receive any Base Salary under Section 4(a) hereof, but shall continue to participate in Company benefit plans in accordance with Section 4(d) and the terms of such plans, until the termination of Employee's employment.

(c) By the Company for Cause. The Company may terminate the Employee's employment hereunder for Cause at any time upon written notice to the Employee setting forth in reasonable detail the nature of such Cause.

(d) By the Company Other than for Cause. The Company may terminate the Employee's employment hereunder other than for Cause at any time upon written notice to the Employee.

(e) By the Employee. The Employee may terminate Employee's employment hereunder at any time upon forty-five (45) days' notice to the Company. In the event of termination of the Employee pursuant to this Section 5(e), the Company may elect to waive the period of notice, or any portion thereof.

6. Severance Payments and Other Matters Related to Separation from Service.

(a) Final Compensation. Following the termination of the Employee's employment for any reason, the Company shall pay to the Employee: (i) any Base Salary earned but not paid during the final payroll period of the Employee's employment through the date of termination, (ii) pay for any vacation time earned but not used through the date of termination, (iii) any unpaid Discretionary Annual Bonus due to Employee for the calendar year prior to the year in which the termination occurs, and (iv) any business expenses incurred by the Employee but unreimbursed on the date of termination, provided that such expenses and required substantiation and documentation are submitted within thirty (30) days of termination and that such expenses are reimbursable under Company policy (all of the foregoing, "Final Compensation"). Any Base Salary and any earned, unused vacation time shall be paid to the Employee at the time required by law, but not later than the Company's next regular pay date following the date of termination. Any reimbursable business expenses shall be paid within sixty (60) days following the date that the Employee submits such expenses to the Company. Other than as expressly provided in Section 6(b), the Company shall have no further obligation to the Employee hereunder.

(b) Severance. In the event the Employee's employment terminates pursuant to Section 5(a), 5(b) or 5(d) of this Agreement, in addition to Final Compensation, (i) the vesting of all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time then held by Employee (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this Agreement) shall accelerate by twelve (12) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) the Company shall pay the Employee (A) a lump sum equal to the Base Salary divided by twelve (12), then multiplied by the number of months of the Severance Period (as defined below) (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below), and (C) an additional one-time bonus of $16,500 (such payment, the "One-Time Bonus"). Subject to Sections 6(d) and 7(a) of this Agreement (x) the Severance Payment and the One-Time Bonus shall be paid by the sixtieth (60th) day following the date of termination and (y) the Post-Termination Bonus shall be paid at or around the time that annual bonuses are paid to other similarly situated employees of the Company, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Parent's 2018 Incentive Award Plan, the Parent's 2015 Equity Incentive Plan or any other equity plan, the Employee shall be immediately 100% fully vested in all unvested equity awards of Parent or its Affiliates that vest solely based on the passage of time (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Employee either before or after the date of this

Agreement and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement). The "Severance Period" shall be nine (9) months; provided, that if the Employee's separation from service occurs during the twelve (12) months following a Change in Control, then the Severance Period shall be twelve (12) months.

(c) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the calendar year in which the termination occurs; provided that if the termination occurs in the twelve (12) month period following a Change in Control, the Post-Termination Bonus shall be equal to the Target Bonus for the calendar year in which such termination occurs.

(d) Release of Claims. The Employee's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Employee's signing and returning to the Company (and not revoking) a general release of claims in the form provided by the Company at the time the Employee's employment is terminated (the "Employee Release"). The Employee must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein.

(e) Effect of Termination. Payment by the Company of Final Compensation and the payments and benefits set forth in Section 6(b) shall constitute the sole obligations of the Company in connection with the termination of the Employee's employment hereunder. Except for any right of the Employee to continue medical and dental plan participation in accordance with applicable law, benefits shall terminate pursuant to the terms of the applicable benefit plans based on the date of termination of the Employee's employment without regard to any of the payments set forth in Section 6(b).

(f) Survival. Provisions of this Agreement shall survive any termination if so provided herein or if necessary or desirable to accomplish the purposes of other surviving provisions, including without limitation the obligations of the Employee under Section 8 hereof. The obligation of the Company to make, and the right of the Employee to retain, any payments or benefits set forth in Section 6(b) is expressly conditioned upon the Employee's continued full performance of obligations under Section 8 and the Confidential Information and Non-Competition Agreement.

7. Timing of Payments and Section 409A.

(a) Notwithstanding anything to the contrary in this Agreement, if at the time of the Employee's termination of employment, the Employee is a Specified Employee (as defined below), such amounts that may be subject to the Specified Employee rules set forth at (a)(2)(B)(i) of Section 409A of the Code ("Section 409A") and payable under Section 6 on account of such Separation from Service (as defined below) that would (but for this provision) be payable within six (6) months following the date of termination, shall instead be paid on the next business day following the expiration of such six (6) month period.

(b) For purposes of this Agreement, "Separation from Service" shall be determined in a manner consistent with subsection (a)(2)(A)(i) of Section 409A, and the term "Specified Employee" shall mean an individual determined by the Company to be a specified employee as defined in subsection (a)(2)(B)(i) of Section 409A.

(c) Each payment made under this Agreement shall be treated as a separate payment and the right to a series of installment payments under this Agreement is to be treated as a right to a series of separate payments.

(d) The Employee's right to reimbursement for business expenses hereunder shall be subject to the following additional rules: (i) the amount of expenses eligible for reimbursement during any calendar year shall not affect the expenses eligible for reimbursement in any other taxable year, (ii) reimbursement shall be made not later than December 31 of the calendar year following the calendar year in which the expense was incurred, and (iii) the right to reimbursement is not subject to liquidation or exchange for any other benefit.

(e) In no event shall the Company have any liability relating to any payment or benefit under this Agreement failing to comply with, or be exempt from, the requirements of Section 409A.

8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Employee's employment with the Company, the Employee has executed the Confidential Information and Non-Competition Agreement, which the Company and Employee acknowledge and agree shall be considered a separate contract. In addition, Employee represents and warrants that Employee shall be able to and/or will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Employee's employment, Employee shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Employee; provided that, the Employee will not have an obligation under this paragraph with respect to any claim that the Employee has filed directly against the Company or related persons or entities. The Employee's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Employee's employment, Employee also shall reasonably cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Employee was employed by the Company, provided Employee will not have any obligation under this paragraph with respect to any claim that Employee has filed directly against the Company or related persons or entities. The Company shall reimburse Employee for any reasonable out-of-pocket expenses incurred in connection with Employee's performance of obligations pursuant to this Section 8(b).

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section

4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

(b) Notwithstanding any provision of Section 9(a) to the contrary, if the Reduction Method or the Pro Rata Reduction Method would result in any portion of the Payment being subject to taxes pursuant to Section 409A that would not otherwise be subject to taxes pursuant to Section 409A, then the Reduction Method and/or the Pro Rata Reduction Method, as the case may be, shall be modified so as to avoid the imposition of taxes pursuant to Section 409A as follows: (i) as a first priority, the modification shall preserve to the greatest extent possible, the greatest economic benefit for Employee as determined on an after-tax basis; (ii) as a second priority, Payments that are contingent on future events (*e.g.*, being terminated without Cause), shall be reduced (or eliminated) before Payments that are not contingent on future events; and (iii) as a third priority, Payments that are "deferred compensation" within the meaning of Section 409A shall be reduced (or eliminated) before Payments that are not deferred compensation within the meaning of Section 409A.

(c) Unless Employee and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Employee and the Company within fifteen (15) calendar days after the date on which Employee's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Employee or the Company) or such other time as requested by Employee or the Company.

(d) If Employee receives a Payment for which the Reduced Amount was determined pursuant to clause (x) of Section 9(a) and the Internal Revenue Service determines thereafter that some portion of the Payment is subject to the Excise Tax, Employee agrees to promptly return to the Company a sufficient amount of the Payment (after reduction pursuant to clause (x) of Section 9(a)) so that no portion of the remaining Payment is subject to the Excise Tax. For the avoidance of doubt, if the Reduced Amount was determined pursuant to clause (y) of Section 9(a), Employee shall have no obligation to return any portion of the Payment pursuant to the preceding sentence.

(e) Notwithstanding anything contained herein to the contrary, the requirements of this Section 9 shall apply only to the extent the Company has completed an "initial public offering" which results in the Company's stock being publicly traded on an applicable public exchange.

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company.

11. Withholding. All payments made by the Company under this Agreement shall be reduced by any tax or other amounts required to be withheld by the Company under applicable law.

12. Assignment.

(a) Neither the Company nor the Employee may make any assignment of this Agreement or any interest herein, by operation of law or otherwise, without the prior written consent of the other; provided, however, that the Company may assign its rights and obligations under this Agreement without the consent of the Employee in the event that (i) the Employee is transferred to a position with any of the Affiliates or (ii) the Company shall hereafter effect a reorganization, consolidate with, or merge into, any Person or transfer all or substantially all of its properties or assets to any Person. This Agreement shall inure to the benefit of and be binding upon the Company and the Employee, their respective successors, executors, administrators, heirs and permitted assigns.

(b) The Company will require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business and/or assets of the Company to assume expressly and agree to perform this Agreement in the same manner and to the same extent that the Company would be required to perform it if no such succession had taken place. As used in this Agreement, "Company" shall mean the Company as hereinbefore defined and any successor to its business and/or assets as aforesaid.

13. Severability. If any covenants or such other provisions of this Agreement are found to be invalid or unenforceable by a final determination of a court of competent jurisdiction, (a) the remaining terms and provisions hereof shall be unimpaired, and (b) the invalid or unenforceable term or provision hereof shall be deemed replaced by a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision hereof.

14. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of either party to require the performance of any term or obligation of this Agreement, or the waiver by either party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Employee at Employee's last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice

so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Confidential Information and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement, together with the Confidential Information and Non-Competition Agreement, supersedes all prior negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee as well as the Change in Control Agreement between the Company and the Employee effective as of September 15, 2018) relating to the subject matter of this Agreement. For clarity, the definition of "Cause" in Section 4(e) of the Confidential Information and Non-Competition Agreement shall only be applicable to that section, whereas the definition of Cause in Section 1(b) of this Agreement shall be applicable throughout this Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that options and other equity awards may be granted to Employee under and pursuant to the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by an expressly authorized representative of the Company.

18. Headings. The headings and captions in this Agreement are for convenience only and in no way define or describe the scope or content of any provision of this Agreement.

19. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be an original and all of which together shall constitute one and the same instrument.

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof. The Company and Employee agree that any dispute concerning this Agreement shall be heard exclusively by a court of competent jurisdiction within the Commonwealth of Massachusetts. By signing below, Employee acknowledges that Employee is subject to the personal jurisdiction of the Massachusetts courts in any county where the Company has operations or facilities. The Employee and Company further agree that any such dispute shall be tried by a judge alone, and they hereby waive and forever renounce the right to a trial before a civil jury in any such dispute.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, this Agreement has been executed as a sealed instrument by the Company, by its duly authorized representative, and by the Employee, as of the date first above written.

EMPLOYEE	**KINIKSA PHARMACEUTICALS CORP.**
/s/ Mark Ragosa	By: /s/ Thomas Beetham
Name: Mark Ragosa	Name: Thomas Beetham
	Title: EVP, Chief Legal Officer

KINIKSA PHARMACEUTICALS, LTD.

NON-EMPLOYEE DIRECTOR COMPENSATION PROGRAM

Non-employee members of the board of directors (the "***Board***") of Kiniksa Pharmaceuticals, Ltd. (the "***Company***") shall receive cash and equity compensation as set forth in this Non-Employee Director Compensation Program (as amended effective as of April 9, 2021, this "***Program***"). The cash and equity compensation described in this Program shall be paid or be made, as applicable, automatically and without further action of the Board, to each member of the Board who is not an employee of the Company or any parent or subsidiary of the Company (each, a "***Non-Employee Director***") who is entitled to receive such cash or equity compensation, unless such Non-Employee Director declines the receipt of such cash or equity compensation by written notice to the Company. This Program shall remain in effect until it is revised or rescinded by further action of the Board. This Program may be amended, modified or terminated by the Board at any time in its sole discretion. The terms and conditions of this Program shall supersede any prior cash and/or equity compensation arrangements for service as a member of the Board between the Company and any of its Non-Employee Directors. No Non-Employee Director shall have any rights hereunder.

I. CASH COMPENSATION

A. Annual Retainers. Each Non-Employee Director shall receive an annual retainer of $40,000 for service on the Board.

B. Additional Annual Retainers. In addition, each Non-Employee Director shall receive the following annual retainers:

1. *Chairman of the Board or Lead Independent Director*. A Non-Employee Director serving as Chairman of the Board or Lead Independent Director shall receive an additional annual retainer of $30,000 for such service.

2. *Audit Committee*. A Non-Employee Director serving as Chairperson of the Audit Committee shall receive an additional annual retainer of $19,000 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Audit Committee shall receive an additional annual retainer of $9,000 for such service.

3. *Compensation Committee*. A Non-Employee Director serving as Chairperson of the Compensation Committee shall receive an additional annual retainer of $13,400 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Compensation Committee shall receive an additional annual retainer of $6,300 for such service.

4. *Nominating and Corporate Governance Committee*. A Non-Employee Director serving as Chairperson of the Nominating and Corporate Governance Committee shall receive an additional annual retainer of $9,300 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Nominating and Corporate Governance Committee shall receive an additional annual retainer of $5,000 for such service.

5. *Science and Research Committee*. A Non-Employee Director serving as Chairperson of the Science and Research Committee shall receive an additional annual retainer of $13,400 for such service. A Non-Employee Director serving as a member other than the Chairperson of the Science and Research Committee shall receive an additional annual retainer of $6,300 for such service.

C. Payment of Retainers. The annual retainers described in Sections I(A) and I(B) shall be earned on a quarterly basis based on a calendar quarter and shall be paid in cash by the Company in arrears not later than the fifteenth day following the end of each calendar quarter. In the event a Non-Employee Director does not serve as a Non-Employee Director, or in the applicable positions described in Section I(B), for an entire calendar quarter, the retainer paid to such Non-Employee Director shall be prorated for the portion of such calendar quarter actually served as a Non-Employee Director, or in such position, as applicable.

II. EQUITY COMPENSATION

Non-Employee Directors shall be granted the equity awards described below. The awards described below shall be granted under and shall be subject to the terms and provisions of the Company's 2018 Incentive Award Plan or any other applicable Company equity incentive plan then-maintained by the Company (the "***Equity Plan***") and shall be granted subject to award agreements in substantially the form previously approved by the Board. All applicable terms of the Equity Plan apply to this Program as if fully set forth herein, and all grants of share options hereby are subject in all respects to the terms of the Equity Plan and the applicable award agreement.

A. Initial Awards. Each Non-Employee Director who is initially elected or appointed to the Board shall automatically be granted an option to purchase a number of shares equal to $600,000 divided by the Black-Scholes Value (as defined below), rounded down to the nearest whole share, but no more than 80,000 shares, of the Company's Class A common shares on the date of such initial election or appointment. The awards described in this Section II(A) shall be referred to as "***Initial Awards***." No Non-Employee Director shall be granted more than one Initial Award.

B. Subsequent Awards. A Non-Employee Director who (i) has been serving as a Non-Employee Director on the Board for at least six (6) months as of the date of any annual meeting of the Company's shareholders and (ii) will continue to serve as a Non-Employee Director immediately following such meeting, shall be automatically granted an option to purchase a number of shares equal to $300,000 divided by the Black-Scholes Value, rounded down to the nearest whole share, but no more than 40,000 shares, of the Company's Class A common shares on the date of such annual meeting. The awards described in this Section II(B) shall be referred to as "***Subsequent Awards***." For the avoidance of doubt, a Non-Employee Director elected for the first time to the Board at an annual meeting of the Company's shareholders shall only receive an Initial Award in connection with such election, and shall not receive any Subsequent Award on the date of such meeting as well.

C. Black-Scholes Value. For purposes of this Section II, "***Black-Scholes Value***" means the per share fair value of an option calculated on the basis of the Black-Scholes

option pricing model and using as inputs to such model (i) for the value of one Class A common share of the Company, the average closing price of the Company's Class A common shares over each trading day occurring in the 30 calendar days ending on the last day of the month prior to the month in which the grant date of the relevant Initial Award or Subsequent Award occurs and (ii) such other assumptions as are determined by the Company's Chief Accounting Officer on or prior to the grant date of the relevant Initial Award or Subsequent Award.

D. Termination of Employment of Employee Directors. Members of the Board who are employees of the Company or any parent or subsidiary of the Company who subsequently terminate their employment with the Company and any parent or subsidiary of the Company and remain on the Board will not receive an Initial Award pursuant to Section II(A) above, but to the extent that they are otherwise entitled, will receive, after termination of employment with the Company and any parent or subsidiary of the Company, Subsequent Awards as described in Section II(B) above.

E. Terms of Awards Granted to Non-Employee Directors

1. *Exercise Price*. The per share exercise price of each option granted to a Non-Employee Director shall equal the Fair Market Value (as defined in the Equity Plan) of a share of the Company's Class A common shares on the date the option is granted.

2. *Vesting*. Each Initial Award shall vest and become exercisable as to one-third of the shares subject to the Initial Award on the first anniversary of the date of grant and as to the remainder in twenty-four (24) substantially equal monthly installments thereafter, such that the Initial Award shall be fully vested on the third anniversary of the date of grant, subject to the Non-Employee Director continuing in service as a Non-Employee Director through each such vesting date. Each Subsequent Award shall vest and become exercisable in twelve (12) substantially equal monthly installments following the date of grant, subject to the Non-Employee Director continuing in service on the Board as a Non-Employee Director through each such vesting date. Unless the Board otherwise determines, any portion of an Initial Award or Subsequent Award which is unvested or unexercisable at the time of a Non-Employee Director's termination of service on the Board as a Non-Employee Director shall be immediately forfeited upon such termination of service and shall not thereafter become vested and exercisable. All of a Non-Employee Director's Initial Awards and Subsequent Awards shall vest in full immediately prior to the occurrence of a Change in Control (as defined in the Equity Plan), to the extent outstanding at such time.

3. *Term*. The maximum term of each share option granted to a Non-Employee Director hereunder shall be ten (10) years from the date the option is granted.

* * * * *

Exhibit 31.1

CERTIFICATIONS

I, Sanj K. Patel, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Kiniksa Pharmaceuticals, Ltd.;

2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;

3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;

4. The registrant's other certifying officer and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:

(a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;

(b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles;

(c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and

(d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and

5. The registrant's other certifying officer and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):

(a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and

(b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

May 6, 2021

/s/ Sanj K. Patel

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

Exhibit 31.2

CERTIFICATIONS

I, Mark Ragosa, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Kiniksa Pharmaceuticals, Ltd.;

2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;

3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;

4. The registrant's other certifying officer and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:

(a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;

(b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles;

(c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and

(d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and

5. The registrant's other certifying officer and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):

(a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and

(b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

May 6, 2021

/s/ Mark Ragosa
Mark Ragosa
Chief Financial Officer
(Principal Financial Officer)

Exhibit 32.1

CERTIFICATION PURSUANT TO
18 U.S.C. SECTION 1350,
AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002

I, Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Directors of Kiniksa Pharmaceuticals, Ltd. (the "Company"), hereby certify, pursuant to 18 U.S.C. §1350, as adopted pursuant to §906 of the Sarbanes-Oxley Act of 2002, that, to the best of my knowledge:

(1) The Quarterly Report on Form 10-Q of the Company for the period ended March 31, 2021 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and

(2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

May 6, 2021

/s/ Sanj K. Patel
Sanj K. Patel
Chief Executive Officer and Chairman of the Board of Directors
(Principal Executive Officer)

Exhibit 32.2

CERTIFICATION PURSUANT TO
18 U.S.C. SECTION 1350,
AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002

I, Mark Ragosa, Chief Financial Officer of Kiniksa Pharmaceuticals, Ltd. (the "Company"), hereby certify, pursuant to 18 U.S.C. §1350, as adopted pursuant to §906 of the Sarbanes-Oxley Act of 2002, that, to the best of my knowledge:

(1) The Quarterly Report on Form 10-Q of the Company for the period ended March 31, 2021 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and

(2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

May 6, 2021

/s/ Mark Ragosa
Mark Ragosa
Chief Financial Officer
(Principal Financial Officer)