

**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, 2020  
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**  
(State or Other Jurisdiction of  
Incorporation or Organization)

**98-1327726**  
(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	<input type="checkbox"/>	Accelerated Filer	<input checked="" type="checkbox"/>
Non-accelerated Filer	<input type="checkbox"/>	Smaller Reporting Company	<input checked="" type="checkbox"/>
		Emerging Growth Company	<input checked="" type="checkbox"/>

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, 2020, there were 55,801,712 common shares outstanding in aggregate, comprised of:

22,285,129 Class A common shares, par value \$0.000273235 per share

2,463,011 Class B common shares, par value \$0.000273235 per share

14,995,954 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, 2020**

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## 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 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 of the novel coronavirus 2019, or COVID-19, pandemic on our business, including our clinical trials and operations;
- our status as a clinical-stage biopharmaceutical company and our expectation to incur losses in the future;
- our future capital needs and our need to raise additional funds;
- our limited operating history;
- the lengthy and expensive clinical development process with its uncertain outcome and potential for clinical failure or delay, including due to the COVID-19 pandemic;
- the decision by any applicable regulatory authority whether to clear our product candidates for clinical development and, ultimately, whether to approve them for marketing and sale;
- our ability to anticipate and prevent adverse events caused by our product candidates;
- our ability to identify, in-license, acquire, discover or develop additional product candidates;
- our ability to have our product candidates manufactured in accordance with regulatory requirements;
- the market acceptance of our product candidates;

- our ability to timely and successfully develop and commercialize our existing 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 our 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 and certain members of senior management 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.

#### **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.

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**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, 2020	December 31, 2019
<b>Assets</b>		
Current assets:		
Cash and cash equivalents	\$ 106,336	\$ 46,928
Short-term investments	97,870	186,452
Prepaid expenses and other current assets	8,849	8,247
Total current assets	213,055	241,627
Property and equipment, net	5,804	6,398
Operating lease right-of-use assets	1,629	1,927
Restricted cash	210	210
Deferred tax assets	5,410	4,372
Total assets	<u>\$ 226,108</u>	<u>\$ 254,534</u>
<b>Liabilities and Shareholders' Equity</b>		
Current liabilities:		
Accounts payable	\$ 4,274	\$ 5,693
Accrued expenses	13,379	20,415
Operating lease liabilities	1,689	1,697
Other current liabilities	—	25
Total current liabilities	19,342	27,830
Non-current liabilities:		
Non-current operating lease liabilities	551	955
Other long-term liabilities	381	326
Total liabilities	20,274	29,111
Commitments and contingencies (Note 12)		
Shareholders' equity:		
Class A common shares, par value of \$0.000273235 per share; 21,519,500 shares and 19,245,201 shares issued and outstanding as of March 31, 2020 and December 31, 2019, respectively	6	6
Class B common shares, par value of \$0.000273235 per share; 3,008,373 shares and 4,638,855 shares issued and outstanding as of March 31, 2020 and December 31, 2019	1	1
Class A1 common shares, \$0.000273235 par value; 14,995,954 shares issued and outstanding as of March 31, 2020 and December 31, 2019, respectively	4	4
Class B1 common shares, \$0.000273235 par value; 16,057,618 shares issued and outstanding as of March 31, 2020 and December 31, 2019	4	4
Additional paid-in capital	588,090	581,467
Accumulated other comprehensive income	240	33
Accumulated deficit	(382,511)	(356,092)
Total shareholders' equity	205,834	225,423
Total liabilities and shareholders' equity	<u>\$ 226,108</u>	<u>\$ 254,534</u>

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,	
	2020	2019
Operating expenses:		
Research and development	\$ 20,901	\$ 59,253
General and administrative	8,486	8,394
Total operating expenses	29,387	67,647
Loss from operations	(29,387)	(67,647)
Interest income	789	1,809
Loss before benefit for income taxes	(28,598)	(65,838)
Benefit for income taxes	2,179	17
Net loss	\$ (26,419)	\$ (65,821)
Net loss per share attributable to common shareholders—basic and diluted	\$ (0.48)	\$ (1.27)
Weighted average common shares outstanding—basic and diluted	55,322,690	51,758,353
Comprehensive loss:		
Net loss	\$ (26,419)	\$ (65,821)
Other comprehensive income:		
Unrealized gain on short-term investments and currency translation adjustments, net of tax	207	12
Total other comprehensive income	207	12
Total comprehensive loss	\$ (26,212)	\$ (65,809)

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	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
<b>Balances at December 31, 2019</b>	54,937,628	\$ 15	\$ 581,467	\$ 33	\$ (356,092)	\$ 225,423
Exercise of options	643,867	—	2,414	—	—	2,414
Share-based compensation expense	—	—	4,209	—	—	4,209
Unrealized gain on short-term investments	—	—	—	207	—	207
Net loss	—	—	—	—	(26,419)	(26,419)
<b>Balances at March 31, 2020</b>	<b>55,581,495</b>	<b>\$ 15</b>	<b>\$ 588,090</b>	<b>\$ 240</b>	<b>\$ (382,511)</b>	<b>\$ 205,834</b>

  

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income (Loss)	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
<b>Balances at December 31, 2018</b>	49,489,647	\$ 13	\$ 473,483	\$ (4)	\$ (194,225)	\$ 279,267
Issuance of Class A common shares upon completion of follow-on offering, inclusive of the over-allotment option exercise, net of underwriting discounts and commissions and offering costs	2,816,110	2	48,474	—	—	48,476
Issuance of Class A1 common shares upon completion of private placement, net of underwriting discounts and commissions and offering costs	2,000,000	—	34,511	—	—	34,511
Class A common shares issued or to be issued in connection with the acquisition of all issued and outstanding equity securities of Primatope Therapeutics, Inc.	337,008	—	7,000	—	—	7,000
Exercise of options	50,070	—	181	—	—	181
Share-based compensation expense	—	—	2,893	—	—	2,893
Unrealized gain on short-term investments	—	—	—	12	—	12
Net loss	—	—	—	—	(65,821)	(65,821)
<b>Balances at March 31, 2019</b>	<b>54,692,835</b>	<b>\$ 15</b>	<b>\$ 566,542</b>	<b>\$ 8</b>	<b>\$ (260,046)</b>	<b>\$ 306,519</b>

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,	
	2020	2019
<b>Cash flows from operating activities:</b>		
Net loss	\$ (26,419)	\$ (65,821)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	594	471
Share-based compensation expense	4,209	2,893
Class A common shares issued or to be issued as consideration for Primatope, including milestone payments	—	7,000
Non-cash lease expense	300	334
Accretion of discounts on short-term investments	(228)	(846)
Deferred income taxes	(1,038)	(240)
Changes in operating assets and liabilities:		
Prepaid expenses and other assets	(598)	(1,544)
Accounts payable	(1,197)	(3,343)
Accrued expenses and other liabilities	(7,061)	(376)
Accrued milestones	—	(2,000)
Operating lease liabilities	(413)	(293)
Other long-term liabilities	55	509
Net cash used in operating activities	<u>(31,796)</u>	<u>(63,256)</u>
<b>Cash flows from investing activities:</b>		
Purchases of property and equipment	(223)	(1,561)
Purchases of short-term investments	(26,187)	(181,807)
Proceeds from the maturities of short-term investments	115,200	174,090
Net cash provided by (used in) investing activities	<u>88,790</u>	<u>(9,278)</u>
<b>Cash flows from financing activities:</b>		
Proceeds from issuance of Class A common shares from follow-on offering, net of underwriting commissions and discounts, inclusive of the over-allotment option exercise	—	48,595
Proceeds from issuance of Class A1 common shares from private placement, net of underwriting commissions and discounts	—	34,511
Payments of offering costs	—	(100)
Proceeds from exercise of options and employee share purchase plan	2,414	181
Net cash provided by financing activities	<u>2,414</u>	<u>83,187</u>
<b>Net increase in cash, cash equivalents and restricted cash</b>	<u>59,408</u>	<u>10,653</u>
Cash, cash equivalents and restricted cash at beginning of period	47,138	72,186
Cash, cash equivalents and restricted cash at end of period	<u>\$ 106,546</u>	<u>\$ 82,839</u>
<b>Supplemental information:</b>		
Cash paid for income taxes	\$ —	\$ —
<b>Supplemental disclosure of non-cash investing and financing activities:</b>		
Deferred offering costs included in accrued expenses and accounts payable	\$ —	\$ 19
Property and equipment included in accrued expenses and accounts payable	\$ —	\$ 37

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 has a pipeline of product candidates across various stages of development that are designed to modulate the immunological signaling pathways that are implicated across a spectrum of diseases.

The Company is subject to risks and uncertainties common to early-stage companies in the biopharmaceutical industry and global health, societal, economic and market conditions, including from the impact from the novel coronavirus 2019 (“COVID-19”). There can be no assurance that the Company’s research and development will be successfully completed, that adequate protection for the Company’s technology will be obtained, that any products developed will obtain necessary government regulatory approval or that any products, if approved, will be commercially viable. The Company does not currently generate revenue from sales of any products, and it may never be able to develop or commercialize a marketable product. The Company has not yet successfully completed any Phase 3 or other pivotal clinical trials, obtained any regulatory approvals, manufactured a commercial-scale drug, or conducted sales and marketing activities. 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, or CROs, and contract manufacturing organizations, or CMOs. Even if the Company’s product development efforts are successful, it is uncertain when, if ever, the Company will realize significant revenue from product sales.

***Risk and Uncertainties related to COVID-19***

In addition to risks and uncertainties common to 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, 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. For example, a number of states, including Massachusetts and California where the Company has offices and laboratory space, have issued shelter-in-place or stay-at-home orders for workers in non-essential businesses. The Company’s business is currently considered to be an essential business so, to date, its operations have only been partially affected by this order, temporarily closing access to its California office space and restricted access to its Massachusetts facility to employees that need to be in the office to execute their responsibilities or who work in its research and development laboratory space, with most of the Company’s employees carrying out their responsibilities outside of the office. The Company is also preparing for any potential re-opening of its offices in Massachusetts and California in the event that the shelter-in-place or stay-at-home orders are lifted.

The COVID-19 pandemic, and measures undertaken in response to the pandemic, or the easing of any of such measures, could 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

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

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 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. 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 continued disruptions in the financial markets, which could adversely impact our 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. 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, and the effectiveness of actions taken to contain and treat the disease.

***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") and, Kiniksa Pharmaceuticals (France) SARL ("Kiniksa France"), after elimination of all significant intercompany accounts and transactions.

In assessing the consolidation requirement for variable interest entities ("VIE"s), the Company focuses on identifying whether it has both the power to direct the activities that most significantly impact the VIE's economic performance and the obligation to absorb losses or the right to receive benefits from the VIE. In the event that the Company is the primary beneficiary of a VIE, the assets, liabilities, and results of operations of the VIE would be included in the Company's consolidated financial statements. At March 31, 2020 and during the three months then ended and at December 31, 2019 and during the year then ended, the Company was not the primary beneficiary of a VIE.

***Use of Estimates***

The preparation of the Company's consolidated financial statements in conformity with GAAP requires management to make estimates and assumptions that affect the reported amounts of assets and liabilities, the disclosure of contingent assets and liabilities at the date of the consolidated financial statements and the reported amounts of expenses during the reporting period. Significant estimates and assumptions reflected in these consolidated financial statements include, but are not limited to, the accrual for research and development expenses and the valuation of common shares and share-based awards. Estimates are periodically reviewed in light of changes in circumstances, facts and experience. Changes in estimates are recorded in the period in which they become known. Actual results could differ from those estimates.

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

***Reporting and Functional Currency***

The financial results of the Company's global activities are reported in U.S. dollars ("USD") and its foreign subsidiaries generally utilize 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.

***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, 2020 and the results of its operations for the three months ended March 31, 2020 and 2019 and its cash flows for the three months ended March 31, 2020 and 2019. The results for the three months ended March 31, 2020 are not necessarily indicative of results to be expected for the year ending December 31, 2020, any other interim periods or any future year or period.

***Follow-on Offering and Private Placement***

On February 4, 2019, the Company completed a follow-on offering of 2,654,984 Class A common shares at a public offering price of \$18.26 and a concurrent private placement of 2,000,000 Class A1 common shares at an offering price of \$18.26 per share for aggregate gross proceeds of \$85,000. In addition, on March 1, 2019, the Company completed the sale of 161,126 Class A common shares to the underwriters of the follow-on offering following the exercise in part of their over-allotment option to purchase additional shares at a public offering price of \$18.26 per share for gross proceeds of \$2,942. The aggregate net proceeds to the Company from the follow-on offering and concurrent private placement, inclusive of the over-allotment option exercise, was \$82,988 after deducting underwriting discounts and commissions and other offering costs

***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, 2020, the Company had an accumulated deficit of \$382,511. During the three months ended March 31, 2020, the Company incurred a net loss of \$26,419 and used \$31,796 of cash in operating activities. The Company expects to continue to

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

generate operating losses for the foreseeable future. As of March 31, 2020, the Company had cash, cash equivalents and short-term investments of \$204,206.

Based on its current operating plan, the Company expects that its cash, cash equivalents and short-term investments will be sufficient to fund its operating expenses 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 research and development programs for product candidates, product portfolio expansion or commercialization efforts, 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, 2020 and December 31, 2019, 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 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.

The Company evaluates its short-term investments with unrealized losses for other-than-temporary impairment. When assessing short-term investments for other-than-temporary declines in value, the Company considers such factors as, among other things, how significant the decline in value is as a percentage of the original cost, the Company's ability and intent to retain the investment for a period of time sufficient to allow for any anticipated recovery in fair value and market conditions in general. If any adjustment to fair value reflects a decline in the value of the investment that the Company considers to be "other than temporary," the Company reduces the investment to fair value through a charge to the consolidated statement of operations and comprehensive loss. No such adjustments were necessary during the periods presented.

***Reclassifications***

The Company reclassified certain prior year balances on our consolidated statements of cash flows to conform to current year presentation. Those balances related to prepaid expenses and other assets and non-cash lease expense. These reclassifications had no effect on our net cash used by operating activities or our consolidated statements of operations and comprehensive loss.

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***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, 2020 and December 31, 2019, 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 (see Note 5), the Company maintains a letter of credit for the benefit of the landlord. As of March 31, 2020 and December 31, 2019, the underlying cash balance of \$210 securing this letter of credit, was classified as non-current in its consolidated balance sheet.

***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***

In February 2016, the Financial Accounting Standards Board ("FASB") issued ASU No. 2016-02, *Leases (Topic 842)* ("ASU 2016-02"), which set forth the principles for recognition, measurement, presentation and disclosure of lease arrangements to enhance the transparency and comparability of financial reporting related to the arrangements. ASU 2016-02, including subsequently issued amendments, is collectively referred to as Accounting Standards

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Codification, *Leases (Topic 842)* (“ASC 842”). The Company adopted the standard on January 1, 2019 using the modified retrospective transition approach as applied to leases existing as of the adoption date. The standard is applied to all leases entered into after the initial adoption date.

At the inception of an arrangement, the Company determines whether the arrangement is or contains a “lease” as defined by 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.

In accordance with the guidance in ASU 2016-02, 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.

Although separation of lease and non-lease components is required, certain practical expedients are available. Companies may elect the practical expedient to not separate lease and non-lease components. In which case, the Company would account for each lease component and the related non-lease component together as a single component. 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.

***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 product 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 for goods or services that will be used or rendered for future research and development activities are recorded as prepaid expenses. Such amounts are recognized as an expense as the goods are delivered or the related services are performed, or until it is no longer expected that the goods will be delivered, or the services rendered. Milestone and other payments made to third parties with respect to in-process

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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 evaluating the adequacy of the accrued liabilities, the Company analyzes progress of the studies or clinical trials, including the phase or completion of events, invoices received and contracted costs. Significant judgments and estimates are made in determining the accrued balances at the end of any reporting period. Actual results could differ materially from the Company's estimates. The Company's historical accrual estimates have not been materially different from the actual costs.

***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 8). 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 the historical volatility of publicly traded peer companies and expects to continue to do so until such time as it has adequate historical data regarding the volatility of its own traded share price. The expected term of the Company's options has been determined utilizing the "simplified" method for awards that qualify as "plain-vanilla" options. The expected term of options granted to non-employees is equal to the contractual term of the option award. The risk-free interest rate is determined by reference to the U.S. Treasury yield curve in effect at the time of grant of the award for time periods approximately equal to the expected term of the award. Expected dividend yield is based on the fact that the Company has never paid cash dividends on common shares and does not expect to pay any cash dividends in the foreseeable future.

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.

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***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, 2020 and 2019, 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, 2020 and 2019.

***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 and Kiniksa France, are subject to taxation in their respective countries. Certain of the Company's subsidiaries, primarily Kiniksa US, operate under cost-plus arrangements.

The Company's benefit for income taxes primarily relates to credits generated by its wholly owned subsidiary, Kiniksa US. The benefit for income taxes is a result of 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 tax credits, the Foreign Derived Intangible Income ("FDII") deduction and share-based compensation taxable events. FDII was enacted as part of the tax reform enacted by the United States in December 2017, generally referred to as the U.S. Tax Cuts and Jobs Act with additional proposed regulations and guidance issued in 2019. Upon the completion of the Company's assessment, the Company has included the tax impact of FDII in its income tax expense for the three months ended

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March 31, 2020, based on its understanding of the rules available at the time of this filing. However, the amounts recorded could be impacted in the future when the proposed regulations and guidance are finalized.

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, and uncertain tax positions are treated as discrete items and are recorded in the period in which they arise.

***Recently Adopted Accounting Pronouncements***

In June 2016, the FASB, issued ASU 2016-13, *Financial Instruments: Credit Losses (Topic 326)*, as clarified in ASU 2019-04 and ASU 2019-05. The objective of the standard is to provide information about expected credit losses on financial instruments at each reporting date and to change how other-than-temporary impairments on investment securities are recorded. The standard became effective for the Company beginning on January 1, 2020. The adoption of this standard did not have a material impact on the Company's consolidated financial statements and related disclosures.

In August 2018, the FASB issued ASU 2018-15, *Intangibles—Goodwill and Other— Internal-Use Software (Subtopic 350-40)* (“ASU 2018-15”), which amends ASC 350-40 to address a customer's accounting for implementation costs incurred in a cloud computing arrangement (“CCA”) that is a service contract. ASU 2018-15 aligns the accounting for costs incurred to implement a CCA that is a service contract with the guidance on capitalizing costs associated with developing or obtaining internal-use software. Specifically, the ASU amends ASC 350 to include in its scope implementation costs of a CCA that is a service contract and clarifies that a customer should apply ASC 350-40 to determine which implementation costs should be capitalized in a CCA that is considered a service contract. The standard became effective for the Company beginning on January 1, 2020. The adoption of this standard did not have a material impact on the Company's consolidated financial statements and related disclosures.

***Recently Issued 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 standard will become effective for the Company beginning on January 1, 2021. The Company is currently evaluating the new standard to determine the potential impact of ASU 2019-12 on its consolidated financial statements and related disclosures.

**3. Fair Value of Financial Assets and Liabilities**

Short-term investments as of March 31, 2020 and December 31, 2019 consisted of U.S. Treasury notes which investments were each due within six months of such date. As of March 31, 2020 and December 31, 2019, the fair value of short-term investments was \$97,870 and \$186,452, respectively. As of March 31, 2020, the amortized cost was

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\$97,630 and gross unrealized gain was \$240. As of December 31, 2019, the amortized cost was \$186,415 and gross unrealized gain was \$37.

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, 2020 Using:			
	Level 1	Level 2	Level 3	Total
<b>Assets:</b>				
Restricted cash — money market funds	\$ 210	\$ —	\$ —	\$ 210
Cash equivalents — money market funds	54,855	—	—	54,855
Cash equivalents — U.S. Treasury notes	—	42,047	—	42,047
Short-term investments — U.S. Treasury notes	—	97,870	—	97,870
	<u>\$ 55,065</u>	<u>\$ 139,917</u>	<u>\$ —</u>	<u>\$ 194,982</u>

	Fair Value Measurements as of December 31, 2019 Using:			
	Level 1	Level 2	Level 3	Total
<b>Assets:</b>				
Restricted cash — money market funds	\$ 210	\$ —	\$ —	\$ 210
Cash equivalents — money market funds	25,207	—	—	25,207
Cash equivalents — U.S. Treasury notes	—	10,192	—	10,192
Short-term investments — U.S. Treasury notes	—	186,452	—	186,452
	<u>\$ 25,417</u>	<u>\$ 196,644</u>	<u>\$ —</u>	<u>\$ 222,061</u>

As of March 31, 2019, the Company did not hold any securities that were in an unrealized loss position. As of December 31, 2019, 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 at December 31, 2019 was \$43,107. 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, 2020 and December 31, 2019.

During the three months ended March 31, 2020 and the year ended December 31, 2019 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, 2020 and December 31, 2019 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.

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**4. Property and Equipment, Net**

Property and equipment, net consisted of the following:

	March 31, 2020	December 31, 2019
Furniture, fixtures and vehicles	\$ 47	\$ 47
Computer hardware and software	344	344
Leasehold improvements	3,627	3,627
Lab equipment	4,705	4,685
Construction in progress	—	20
Total property and equipment	8,723	8,723
Less: Accumulated depreciation	(2,919)	(2,325)
Total property and equipment, net	<u>\$ 5,804</u>	<u>\$ 6,398</u>

Depreciation expense was \$594 and \$471 during the three months ended March 31, 2020 and 2019, respectively.

**5. Leases**

Kiniksa US leases office and laboratory space under operating leases. Leases with an initial term of 12 months or less are not recorded on the balance sheet; the Company recognizes lease expense for these leases on a straight-line basis over the lease term. For lease agreements entered into or reassessed after the Company's adoption of ASC 842, the Company will combine lease and non-lease components. Kiniksa US's leases have remaining lease terms of less than 2 years.

On March 13, 2018, Kiniksa US entered into an operating lease in Lexington, Massachusetts for office and laboratory space that comprises the headquarters for Kiniksa US and on June 26, 2018, Kiniksa US entered into an amendment to the lease expanding the rentable space to a total of 27,244 square feet. On November 7, 2018, Kiniksa US entered into an amendment (the "Third Amendment") to the lease expanding the rentable space to a total of 55,924 square feet which were occupied in phases through December 2019. The lease expires on July 31, 2021. Monthly lease payments include base rent, as well as, ancillary charges such as the share of operating expenses and real estate taxes. Base rent under the Third Amendment is \$138 per month as of December 2019.

On December 21, 2018, Kiniksa US entered into an operating lease in San Diego, California for office space comprising a total of 4,400 square feet. The lease commenced on January 1, 2019 and expires on December 31, 2020. Monthly lease payments for base rent are \$13. Additional fees for ancillary charges such as the share of operating expenses, parking and real estate taxes are not included in the base rent.

The components of lease cost consisted of operating lease costs and variable lease costs were \$340 and \$50 for the three months ended March 31, 2020, respectively. The components of lease cost consisted of operating lease costs and variable lease costs were \$401 and \$22 for the three months ended March 31, 2019, respectively. As of March 31, 2020, the weighted-average lease term was 1.22 years and the discount rate was 7.16%.

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Maturities of operating leases liabilities were as follows:

<b>As of March 31,</b>	
2020	\$ 1,366
2021	972
2022	—
2023 and thereafter	—
Total future minimum lease payments	\$ 2,338
Less imputed interest	(98)
Present value of lease liabilities	\$ 2,240

**6. Accrued Expenses**

Accrued expenses consisted of the following:

	<b>March 31, 2020</b>	<b>December 31, 2019</b>
Accrued research and development expenses	\$ 8,479	\$ 11,813
Accrued employee compensation and benefits	3,456	7,089
Accrued legal and professional fees	726	1,087
Other	718	426
	<u>\$ 13,379</u>	<u>\$ 20,415</u>

**7. Common Shares**

On February 4, 2019, the Company completed a follow-on offering of 2,654,984 Class A common shares at a public offering price of \$18.26 and a concurrent private placement of 2,000,000 Class A1 common shares at an offering price of \$18.26 per share for aggregate gross proceeds of \$85,000. In addition, on March 1, 2019, the Company completed the sale of 161,126 Class A common shares to the underwriters of the follow-on offering following the exercise in part of their over-allotment option to purchase additional shares at a public offering price of \$18.26 per share for gross proceeds of \$2,942. The aggregate net proceeds to the Company from the follow-on offering and concurrent private placement, inclusive of the over-allotment option exercise, was \$82,988 after deducting underwriting discounts and commissions 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, 2020, 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.

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*Dividends*

Common shareholders are entitled to receive dividends, as may be declared by the Company's board of directors. Through March 31, 2020, 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.

**8. 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").

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 (i) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (ii) a smaller number of Class A common shares determined by the Company's board of directors. In December 2019, the board of directors approved the automatic increase as of January 1, 2020 of 2,197,505 shares, equal to 4% of the as-converted Class A common shares outstanding on December 31, 2019. 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, lapse unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited under the 2018 Plan or the 2015 Plan will be added back to the Class A common shares available for issuance under the 2018 Plan. As of March 31, 2020, 3,226,660 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 such awards and the

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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.

As of March 31, 2020, there were 3,477,438 Class A common shares subject to outstanding awards under the 2015 Plan and reserved for issuance under the 2015 Plan pursuant to such awards. On May 23, 2018, the effective date of the 2018 Plan, the Company ceased granting awards under the 2015 Plan and no Class A common shares were available for future grant under the 2015 Plan in connection with the 2018 Plan becoming effective. However, 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 are forfeited, lapse unexercised or are settled in cash become available for issuance under the 2018 Plan.

The exercise price for incentive share options was determined by the Company's board of directors. All incentive share options granted to any person possessing 10% or less of the total combined voting power of all classes of shares could not have an exercise price of less than 100% of the fair market value of the Class A common shares on the grant date. All incentive share options granted to any person possessing more than 10% of the total combined voting power of all classes of shares could not have an exercise price of less than 110% of the fair market value of the Class A common shares on the grant date. The option term for incentive share options could not be greater than 10 years. Incentive share options granted to persons possessing more than 10% of the total combined voting power of all classes of shares could not have an option term of greater than five years. The vesting period for equity-based awards was determined by the board of directors, which was generally four to six years. For awards granted to employees and non-employees with four-year vesting terms, 25% of the option vests on the first anniversary of the grant date and the remaining shares vest equally each month for three years thereafter. For awards granted to employees with six-year vesting terms, 16% of the option vests on the first anniversary of the grant date and the remaining shares vest based on a predetermined vesting schedule for five years thereafter.

***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 (i) 1% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (ii) a smaller number of Class A common shares determined by the Company's board of directors, provided that no more than 6,420,000 Class A common shares may be issued under the 2018 ESPP. As of March 31, 2020, 598,334 Class A common shares were available for future issuance under the 2018 ESPP. In December 2019, the Company's board of directors determined that the January 1, 2020 automatic increase in shares available under the 2018 ESPP would be zero shares.

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**NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)**  
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**Options**

Share option activity under the Plans is summarized as follows:

	Number of Shares	Weighted Average Fair Value
Outstanding as of December 31, 2019	8,491,734	\$ 7.68
Granted	1,455,250	\$ 10.74
Exercised	(643,867)	\$ 2.50
Forfeited	(234,919)	\$ 10.89
Cancelled	(77,326)	\$ 16.72
Outstanding as of March 31, 2020	8,990,872	\$ 8.39
Options exercisable as of March 31, 2020	3,176,450	\$ 8.26
Options unvested as of March 31, 2020	5,814,422	\$ 9.63

**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, 2020 and 2019 were as follows, presented on a weighted-average basis:

	Three Months Ended March 31,	
	2020	2019
Risk-free interest rate	0.82 %	2.58 %
Expected term (in years)	6.25	6.25
Expected volatility	79.83 %	78.18 %
Expected dividend yield	0 %	0 %

**Riloncept Long-Term Incentive Plan**

In December 2019, the compensation committee of the Company's board of directors approved the Company's Riloncept Long-Term Incentive Plan ("RLTIP") under the 2018 Plan to incentivize eligible employees of the Company or any of its subsidiaries to achieve U.S. Food and Drug Administration ("FDA") approval for the commercial sale and marketing of riloncept for the treatment of 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 unit ("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 which the RLTIP Milestone is 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 in accordance with the RLTIP based on the earnout percentage. The number of Class A common shares issuable under the first RSU award ("First RSU Award") as a result of the RLTIP Milestone will be determined in

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accordance with the RLTIIP based on the earnout percentage, and such RSUs will vest on the first anniversary of the Achievement Date, subject to continued employment through such date. The second RSU award will be granted on the Achievement Date with respect to a number of shares determined in accordance with the RLTIIP, 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.

***Restricted Share Units***

An RSU represents the right to receive shares of the Company’s Class A common shares upon vesting of the RSU. The fair value of each RSU is based on the closing price of the Company’s Class A common shares on the date of grant. In December 2019, the Company granted RSUs with service conditions (“Time-Based RSUs”) that vest in one installment on December 31, 2020, subject to the recipient’s continued employment through that date. During the year ended December 31, 2019, the Company also granted the First RSU Award as part of the RLTIIP, which becomes eligible to vest upon the Achievement Date and will vest upon the first anniversary of such date, subject to the recipient’s continued employment through that date. In the event the RLTIIP Milestone is not achieved, none of the First RSU Award will vest.

The grant-date fair value of the Time-Based RSUs granted in 2019 was \$1,250 and will be recognized on a straight-line basis through the vest date for these RSUs. For the three months ended March 31, 2020, the Company recognized \$261 in Time-Based RSU expense. The grant-date fair value of the First RSU Award was \$2,994 and will be recognized when the RLTIIP Milestone is deemed probable of achievement through the date the First RSU Award will vest. During the three months ended March 31, 2020, the Company did not recognize compensation expense related to the First RSU Award, as achievement of the RLTIIP Milestone was determined to be not probable.

The following table summarizes RSU activity for the three months ended March 31, 2020:

	Number of Shares	Weighted Average Grant Date Fair Value
Unvested RSUs as of December 31, 2019	328,296	\$ 12.93
Granted	—	\$ —
Vested	—	\$ —
Forfeited	(26,253)	\$ 12.93
Unvested RSUs as of March 31, 2020	<u>302,043</u>	<u>\$ 12.93</u>

***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,	
	2020	2019
Research and development expenses	\$ 1,769	\$ 1,187
General and administrative expenses	2,440	1,706
	<u>\$ 4,209</u>	<u>\$ 2,893</u>

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**9. License and Acquisition Agreements**

***Biogen Asset Purchase Agreement***

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to vixarelimab (KPL-716) and other antibodies covered by certain patent rights, including patents and other intellectual property rights, clinical data, know-how, and clinical drug supply. In addition, Biogen granted to the Company a non-exclusive, sublicensable, worldwide license to certain background patent rights related to the 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. No milestones were achieved or paid during the three months ended March 31, 2020. 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, 2020, the Company recorded research and development expense of \$64, primarily related to a milestone and the annual maintenance fee both in connection with the retained contracts. During the three months ended March 31, 2019, the Company recorded research and development expense of \$10,002 primarily related to an accrued milestone associated with the achievement of a specified clinical milestone event due under the Biogen Agreement.

***Novo Nordisk License Agreement***

In August 2017, the Company entered into a license agreement (the "Novo Nordisk Agreement") with Novo Nordisk A/S ("Novo Nordisk"), pursuant to which the Company was granted an exclusive, sublicensable, worldwide license under certain intellectual property rights controlled by Novo Nordisk to make, use, develop and commercialize KPL-045 for all indications.

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**NOTES TO CONSOLIDATED FINANCIAL STATEMENTS (Continued)**  
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In consideration for the license, the Company made an upfront payment of \$1,500 to Novo Nordisk. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

In January 2020, the Company terminated the Novo Nordisk Agreement in accordance with the terms and conditions of the agreement.

The Company did not record any research and development expense during the three months ended March 31, 2020 and 2019, in connection with milestone payments due under the Novo Nordisk Agreement.

***Primatope Stock Purchase Option Agreement***

In September 2017, the Company entered into a stock purchase option agreement (the “Primatope Agreement”) with Primatope Therapeutics, Inc. (“Primatope”), pursuant to which the Company was granted a license to certain intellectual property rights owned or controlled by Primatope to research, develop, and manufacture the preclinical antibody, KPL-404.

The agreement provided the Company with an exclusive call option to purchase 100% of the equity securities of Primatope. Upon execution of the agreement, the Company made \$500 in upfront payments for the initial option period through April 2018 (the “Initial Option Period”). The Primatope Agreement allowed for up to three extensions of the Initial Option Period through January 2019 (including the initial option period, the “Option Period”) for total extension payments of up to \$800. Through December 31, 2018, the Company made payments totaling \$800 to extend the Option Period to January 15, 2019. During the Option Period, the Company could conduct research and preclinical work to assess the viability of the asset.

The Company determined that the call option represented a variable interest in Primatope and that Primatope is a VIE. However, as the Company had no ability to control the board of directors or direct the ongoing activities of Primatope during the Option Period, the Company did not have power over the activities that most significantly impact Primatope’s economic performance and was not the primary beneficiary of Primatope. As a result, the Company did not consolidate the assets, liabilities, and results of operations of Primatope.

In January 2019, the Company exercised the call option and in March 2019, the Company acquired all of the issued and outstanding equity securities of Primatope (the “Primatope Acquisition”) in exchange for \$18,000 comprised of upfront consideration of \$10,000 at closing and milestone payments of \$5,000, which had been achieved as of the closing date, and in June 2019, the Company made the final milestone payment of \$3,000, which was achieved during the year ended December 31, 2019 following the closing, each paid in a combination of cash and Class A common shares (inclusive of escrow and holdback amounts) in accordance with the Primatope Agreement. At the closing of the Primatope Acquisition, Primatope became a wholly owned subsidiary of the Company and the acquisition was accounted for as an asset acquisition as it did not meet the definition of a business. The Company recorded the upfront payment and milestone payments 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.

During the three months ended March 31, 2020, the Company did not incur any research and development expense directly in connection with milestone or other payments related to the Primatope Acquisition or the Primatope Agreement. During the three months ended March 31, 2019, the Company recorded research and development expense of \$18,000 related to the Primatope Acquisition.

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***Beth Israel Deaconess Medical Center License Agreement***

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.

The Company did not incur any research and development expense in connection with the BIDMC Agreement during the three months ended March 31, 2020 and 2019.

***Regeneron License Agreement***

In September 2017, the Company entered into a license agreement (the “Regeneron Agreement”) with Regeneron Pharmaceuticals, Inc. (“Regeneron”), pursuant to which the Company has been granted an exclusive, sublicensable license under certain intellectual property rights controlled by Regeneron to develop and commercialize rilonacept in certain fields and territories. The Company is obligated to use commercially reasonable efforts to develop and commercialize such licensed products.

In exchange for these rights, the Company made an upfront payment of \$5,000. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

Under the Regeneron Agreement, the Company is also obligated to make payments to Regeneron of up to an aggregate of \$27,500 upon the achievement of specified regulatory milestones. Upon commercialization of the licensed products, the parties will share profits equally for all commercial products, after deducting certain commercialization expenses subject to specified limits.

Under the Regeneron Agreement, the Company is solely responsible for all development and commercialization activities and costs in its territories. The Company is also responsible for costs related to the filing, prosecution and maintenance of certain licensed patent rights.

The parties also entered into a clinical supply agreement under which Regeneron agreed to manufacture the developed product during the clinical phase. During the three months ended March 31, 2020, the Company did not incur any research and development expense related to the purchase of drug materials under this agreement. During the three months ended March 31, 2019, the Company recorded research and development expense of \$515, related to the purchase of drug materials under this agreement. As of March 31, 2020 and December 31, 2019, the Company had non-cancelable purchase commitments under the clinical supply agreement (see Note 12).

The Regeneron Agreement will expire when the Company is no longer developing or commercializing any licensed product under the Regeneron Agreement. Either party may terminate the agreement upon the other party’s insolvency or bankruptcy or for material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches). Regeneron has the right to terminate the agreement if the Company suspends its development or commercialization activities for a consecutive 12-month period or does not grant a sublicense to a

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third-party to perform such activities, or if the Company challenges any of the licensed patent rights. The Company may terminate the agreement at any time that is 18 months after the effective date of the agreement with 180 days' written notice or with one year's written notice if the Company terminates the agreement following U.S. marketing approval of a riloncept product developed by the Company. The Company may also terminate the agreement with three months' written notice if the products are determined to have certain safety concerns.

The Company did not incur any research and development expense directly related to milestone payments due under the Regeneron Agreement during the three months ended March 31, 2020 and 2019.

***MedImmune License Agreement***

In December 2017, the Company entered into a license agreement (the "MedImmune Agreement") with MedImmune, Limited ("MedImmune"), pursuant to which MedImmune granted the Company an exclusive, sublicensable, worldwide license to certain intellectual property rights to make, use, develop and commercialize mavrilimumab. Under the MedImmune Agreement, the Company also acquired reference rights to relevant manufacturing and regulatory documents and MedImmune's existing supply of mavrilimumab drug substance and product. The Company is obligated 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 met 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. As of December 31, 2018 and 2017, the Company determined that the payment related to this milestone was probable and, therefore, recognized research and development expense and an accrued milestone of \$10,000 during the year ended December 31, 2017. 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. 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.

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During the three months ended March 31, 2020 and 2019, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

***Kite Clinical Collaboration Agreement***

In December 2019, the Company entered into a clinical collaboration (the “Kite Agreement”) with Kite Pharma, Inc., a Gilead Company (“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. The objective of the Phase 2 trial is to determine the effect of mavrilimumab on the safety of Yescarta. Treatment related induction of GM-CSF has been identified through clinical, translational and preclinical studies as a potential key signal associated with side effects of chimeric antigen receptor T, or CAR T, cell therapy. Preclinical evidence suggest the potential for interruption of GM-CSF signaling to disrupt CAR T cell mediated inflammation without disrupting anti-tumor activity. Kite will be the sponsor of this study and responsible for its conduct. Under the Kite Agreement, Kite and the Company agreed to share a portion of the costs incurred in conducting the study.

The Kite Agreement will expire on the one-year anniversary of the date that Kite provides the clinical study report to the Company or unless otherwise terminated in accordance with the Kite Agreement.

The Company evaluated the agreement and determined all costs and expenses will be recognized as incurred as research and development expenses. During the three months ended March 31, 2020, the Company did not record any research and development expense in connection with the Kite Agreement.

**10. 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 7). As the liquidation and dividend rights are identical, losses are allocated on a proportionate basis and the resulting net loss per share attributed to common shareholders will, therefore, be the same for both Class A and Class B common shares on an individual or combined basis.

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

	Three Months Ended March 31,	
	2020	2019
<b>Numerator:</b>		
Net loss attributable to common shareholders	\$ (26,419)	\$ (65,821)
<b>Denominator:</b>		
Weighted average common shares outstanding—basic and diluted	55,322,690	51,758,353
Net loss per share attributable to common shareholders— basic and diluted	\$ (0.48)	\$ (1.27)

The Company’s unvested restricted shares and 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, unvested restricted shares 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

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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,	
	2020	2019
Options to purchase common shares	8,990,872	7,195,501
Unvested restricted shares	—	609,593
Unvested RSUs	302,043	—
	<u>9,292,915</u>	<u>7,805,094</u>

**11. 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 and Kiniksa France, 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, 2020 varied from the Bermuda statutory rate of zero primarily due to income subject to United States taxation under the Kiniksa US cost-plus arrangement with the Company, net of the deduction for FDII and U.S. federal and state research tax credits. Income tax benefit for the three months ended March 31, 2020 was \$2,179 and includes a net \$1,828 discrete tax benefit primarily due to the tax benefits from share-based compensation taxable events. Income tax benefit for the three months ended March 31, 2019 was not materially different than the Bermuda statutory rate.

As of March 31, 2020, the Company has a deferred tax asset of \$5,410, primarily related to unexercised share-based compensation granted to the employees of Kiniksa US. Management examines all positive and negative evidence to estimate whether sufficient future taxable income in the U.S. will be generated to permit the use of existing deferred tax assets.

**12. 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 9).

***Manufacturing Commitments***

The Company entered into agreements with several contract manufacturing organizations to provide preclinical and clinical trial materials. As of March 31, 2020, the Company had committed to minimum payments under these agreements totaling \$1,508.

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***Rilonacept Long-Term Incentive Plan***

During the year ended December 31, 2019, the Company granted a cash award and the First RSU Award to employees under the RLTIIP. The cash award vests upon the achievement of the RLTIIP Milestone, subject to the recipient's continued employment. The First RSU Award becomes eligible to vest upon the achievement of the RLTIIP Milestone, and will vest upon the first anniversary of such date, subject to the recipient's continued employment through that date. As of March 31, 2020, the Company estimated cash payments of \$1,938 under the RLTIIP. In the event the RLTIIP Milestone is not achieved, the cash award will not be paid and the First RSU Award will not vest.

***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 does not believe that the outcome of any claims under indemnification arrangements will have a material effect on its financial position, results of operations or cash flows, and it has not accrued any liabilities related to such obligations in its consolidated financial statements as of March 31, 2020 or December 31, 2019.

***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, 2019 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 with significant unmet medical need. We have a pipeline of clinical-stage product candidates that 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 signaling pathways that are implicated across a spectrum of diseases. Our product candidates include rilonacept, mavrilimumab, vixarelimab (KPL-716) and KPL-404.

Our lead candidate is rilonacept, an interleukin-1 $\alpha$ , and interleukin-1 $\beta$ , cytokine trap. We are developing rilonacept for the potential treatment of recurrent pericarditis, a painful inflammatory cardiovascular disease with an estimated U.S. prevalent population of approximately 40,000 patients seeking and receiving medical treatment. We are conducting a global, double-blind, placebo-controlled, randomized-withdrawal design, pivotal Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis, named RHAPSODY, for which we have completed enrollment. We expect top-line data from this trial in the second half of 2020. We received Breakthrough Therapy designation from the U.S. Food and Drug Administration, or FDA, for rilonacept for the treatment of recurrent pericarditis in 2019, and reported final data from our open label Phase 2 proof of concept clinical trial of rilonacept in a range of recurrent pericarditis populations at the American Heart Association Scientific Sessions in November 2019.

Mavrilimumab is a monoclonal antibody that antagonizes granulocyte-macrophage colony stimulating factor. 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 are conducting a randomized, double-blind, placebo-controlled, global Phase 2 proof-of-concept trial for the study of mavrilimumab in GCA, for which we have completed enrollment. We expect top-line data from this trial in the second half of 2020. In December 2019, we entered into a clinical collaboration with Kite Pharma, Inc., a Gilead Company, or 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. The objective of the Phase 2 trial is to determine the effect of mavrilimumab on the safety of Yescarta. Treatment related induction of GM-CSF has been identified through clinical, translational and preclinical studies as a potential key signal associated with side effects of chimeric antigen receptor T, or CAR T, cell therapy. Preclinical evidence suggest the potential for interruption of GM-CSF signaling to disrupt CAR T cell mediated inflammation without disrupting anti-tumor activity. Kite will be the sponsor of this study and responsible for its conduct. Further, we recently announced evidence of treatment response with mavrilimumab from an open-label treatment protocol conducted in Italy by an academic investigator in 6 non-mechanically ventilated patients with severe novel coronavirus 2019, or COVID-19, pneumonia and hyperinflammation as well as from an additional 7 such patients who were subsequently treated with mavrilimumab. We have engaged with the FDA and are preparing for a potential registrational development program for mavrilimumab in COVID-19 pneumonia and hyperinflammation. In parallel, academic investigators in the U.S. and Italy are planning investigator-initiated placebo-controlled studies.

Vixarelimab is a monoclonal antibody that simultaneously inhibits the signaling of the cytokines interleukin 31, or IL-31, and oncostatin M, or OSM, by targeting their common receptor subunit, oncostatin M receptor beta, or OSMR $\beta$ . We are conducting a randomized, double-blind, placebo-controlled, Phase 2a clinical trial of vixarelimab in subjects with prurigo nodularis, a chronic inflammatory skin condition with an estimated U.S. prevalence of approximately 300,000 patients. We recently announced data from our Phase 2a trial of vixarelimab in patients with prurigo nodularis, a chronic inflammatory skin condition. The trial met its primary efficacy endpoint: the reduction in weekly-average Worst-Itch Numeric Rating Scale (WI-NRS) from baseline at Week 8 was statistically significantly greater in patients who received vixarelimab versus those who received placebo. We are also conducting an exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus. This randomized, double-blind, placebo-controlled trial is designed to identify chronic pruritic conditions where signaling through OSMR $\beta$  may be playing a role and to investigate the efficacy, safety and tolerability of vixarelimab in reducing the moderate-to-severe pruritus experienced by these subjects. We expect data from cohorts of this trial in the first half of 2020.

KPL-404 is a monoclonal antibody inhibitor of the CD40/CD40L interaction, a central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity. In the first quarter of 2019, we acquired all of the outstanding capital stock of Primatope Therapeutics, Inc., or Primatope, the company that owned or controlled the intellectual property related to KPL-404. In the second half of 2019, we initiated a single-ascending-dose Phase 1 clinical trial of KPL-404 in healthy volunteers. The first-in-human trial is designed to provide safety and pharmacokinetics data as well as data regarding receptor occupancy and T-cell dependent antibody response in these subjects. We expect data in the second half of 2020.

Our future success is dependent on our ability to develop, obtain regulatory approval for and successfully commercialize one or more of our product candidates. We have not yet demonstrated our ability to successfully conduct and complete a Phase 3 or other pivotal clinical trial, obtain regulatory approvals, manufacture a commercial scale drug, or conduct sales and marketing activities. We currently generate no revenue from sales of any products, and we may never be able to develop or commercialize a marketable product.

On February 4, 2019, we completed a follow-on offering of 2,654,984 Class A common shares and concurrent private placement of 2,000,000 Class A1 common shares, both at \$18.26 per share for aggregate gross proceeds of \$85.0 million. In addition, on March 1, 2019, we completed the sale of 161,126 Class A common shares to the underwriters of the follow-on offering following the exercise in part of their over-allotment option to purchase additional shares at \$18.26 per share for gross proceeds of \$2.9 million. The aggregate net proceeds to us from the follow-on offering and concurrent private placement, inclusive of the over-allotment option exercise, was \$83.0 million after deducting underwriting discounts and commissions 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 development and eventual commercialization of one or more of our current or future product candidates. Our net losses were \$26.4 million and \$65.8 million for the three months ended March 31, 2020 and 2019, respectively. As of March 31, 2020, we had an accumulated deficit of \$382.5 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, if we obtain marketing approval for any of our product candidates, we expect to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates. We expect to incur significant expenses in connection with our ongoing activities, particularly as we advance the clinical trials, preclinical activities of our product candidates, and prepare for commercial operations.

As a result, until such time as we can generate significant revenue from product sales, 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 commercialization of one or more of our product candidates or delay our pursuit of potential in-licenses or acquisitions.

Because of the numerous risks and uncertainties associated with product development, 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 generate product sales, we may not become profitable. If we fail to become profitable or are unable to sustain profitability on a continuing basis, then we may be unable to continue our operations at planned levels and be forced to reduce or terminate our operations.

As of March 31, 2020, we had cash, cash equivalents and short-term investments of \$204.2 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***

To date, we have not generated any revenue from any products, and may never be able to develop and commercialize a marketable product. If our development efforts for our product candidates are successful and result in regulatory approval, we may generate revenue in the future from product sales.

### ***Operating Expenses***

#### ***Research and Development Expenses***

Research and development expenses consist primarily of costs incurred in connection with the 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,	
	2020	2019
	(in thousands)	
Rilonacept	\$ 4,037	\$ 5,363
Mavrilimumab	2,210	5,574
Vixarelimab <sup>(1)</sup>	2,655	16,450
KPL-404 <sup>(2)</sup>	1,271	19,426
Unallocated research and development expenses	10,728	12,440
Total research and development expenses	<u>\$ 20,901</u>	<u>\$ 59,253</u>

(1) The amount for the three months ended March 31, 2019 includes expense of \$10.0 million related to a milestone under our asset purchase agreement with Biogen MA, Inc., or Biogen, associated with the achievement of a specified clinical milestone event.

(2) The amount for the three months ended March 31, 2019 includes expense of \$18.0 million related to our acquisition of the issued and outstanding equity securities of Primatope and milestone achievements, paid in a combination of cash and Class A common shares (inclusive of escrow and holdback amounts) in accordance with the stock purchase option agreement with Primatope, or the Primatope Agreement.

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 rilonacept, mavrilimumab, vixarelimab and KPL-404, as well as conduct other preclinical and clinical development including regulatory filings for our 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.

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

- the 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 preclinical development activities, clinical trials and other research and development activities;
- establishing an appropriate safety and efficacy profile with Investigational New Drug, or 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;
- establishing clinical and commercial manufacturing capabilities or making arrangements with third-party manufacturers in order to ensure that we or our third-party manufacturers are able to make product successfully;
- development and timely delivery of clinical-grade and commercial-grade drug formulations that can be used in our clinical trials and for commercial launch;
- obtaining, maintaining, defending and enforcing patent claims and other intellectual property rights;
- significant and changing government regulation;
- launching commercial sales of our product candidates, if and when approved, whether alone or in collaboration with others; and
- maintaining a continued acceptable safety profile of our product candidates following approval, if any, of our product candidates.

#### *General and Administrative Expenses*

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

We expect that our general and administrative expenses will continue to increase in the future as we continue to prepare for potential 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.

#### ***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 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 and Kiniksa Pharmaceuticals (France) SARL are subject to taxation in their respective countries. Our benefit for income taxes relates to credits generated by our wholly owned subsidiary Kiniksa US as well as the tax deductions associated with equity compensation transactions.

As of December 31, 2019, we had federal research and development tax credit carryforwards of approximately \$0.2 million available to reduce future tax liabilities, which begin to expire in 2039. As of December 31, 2019, we had state research and development tax credit carryforwards of approximately \$0.3 million available to reduce future tax liabilities, which begin to expire in 2034.

## Results of Operations

### Comparison of the Three Months Ended March 31, 2020 and 2019

The following table summarizes our results of operations for the three months ended March 31, 2020 and 2019:

	Three Months Ended March 31,		Change
	2020	2019	
	(in thousands)		
Operating expenses:			
Research and development	\$ 20,901	\$ 59,253	\$ (38,352)
General and administrative	8,486	8,394	92
Total operating expenses	<u>29,387</u>	<u>67,647</u>	<u>(38,260)</u>
Loss from operations	(29,387)	(67,647)	38,260
Interest income	789	1,809	(1,020)
Loss before benefit for income taxes	(28,598)	(65,838)	37,240
Benefit for income taxes	2,179	17	2,162
Net loss	<u>\$ (26,419)</u>	<u>\$ (65,821)</u>	<u>\$ 39,402</u>

### Research and Development Expenses

	Three Months Ended March 31,		Change
	2020	2019	
	(in thousands)		
Direct research and development expenses by program:			
Rilonacept	\$ 4,037	\$ 5,363	\$ (1,326)
Mavrilimumab	2,210	5,574	(3,364)
Vixarelimab	2,655	16,450	(13,795)
KPL-404	1,271	19,426	(18,155)
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	7,361	6,424	937
Other	3,367	6,016	(2,649)
Total research and development expenses	<u>\$ 20,901</u>	<u>\$ 59,253</u>	<u>\$ (38,352)</u>

Research and development expenses were \$20.9 million for the three months ended March 31, 2020, compared to \$59.3 million for the three months ended March 31, 2019. The decrease of \$38.4 million was primarily due to expenses incurred in the three months ended March 31, 2019 related to \$10.0 million for a milestone for our vixarelimab program and \$18.0 million for the acquisition of all of the outstanding stock of Primatope.

The direct costs for our rilonacept program were \$4.0 million during the three months ended March 31, 2020, compared to \$5.4 million during the three months ended March 31, 2019, or a decrease of \$1.4 million. During the three months ended March 31, 2020, expenses incurred primarily related to conducting RHAPSODY, our global, pivotal Phase 3 clinical trial in recurrent pericarditis, compared to the three months ended March 31, 2019, in which expenses incurred related to the start of RHAPSODY, including the purchase of clinical drug supply as well as for conducting our open-label Phase 2 proof-of-concept clinical trial.

The direct costs for our mavrilimumab program were \$2.2 million during the three months ended March 31, 2020, compared to \$5.6 million during the three months ended March 31, 2019, or a decrease of \$3.4 million. During the three months ended March 31, 2020, expenses incurred related primarily to clinical trial and supply costs associated with our global Phase 2 clinical trial in GCA compared to the three months ended March 31, 2019, in which expenses incurred related primarily to manufacturing, process development, and the start of our GCA clinical trial.

The direct costs for our vixarelimab program were \$2.7 million during the three months ended March 31, 2020, compared to \$16.5 million during the three months ended March 31, 2019, or a decrease of \$13.8 million. During the three months ended March 31, 2020, expenses incurred primarily related to expenses incurred for our Phase 2a clinical trial in prurigo nodularis and our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus, including manufacturing and development costs for our clinical drug supply, compared to the three months ended March 31, 2019, in which expenses incurred related to an accrued milestone of \$10.0 million under our asset purchase agreement with Biogen associated with the achievement of a specified clinical milestone event as well as expenses incurred for our Phase 2a clinical trial in prurigo nodularis including manufacturing and development costs for our clinical drug supply.

The direct costs for our KPL-404 program were \$1.3 million during the three months ended March 31, 2020, compared to \$19.4 million during the three months ended March 31, 2019, or a decrease of \$18.1 million. During the three months ended March 31, 2020, expenses incurred primarily related to preclinical expenses and initiation of our Phase 1 trial. During the three months ended March 31, 2019, expenses incurred related to \$18.0 million of expense related to the acquisition of all the issued and outstanding equity securities of Primatope in accordance with the terms and conditions of the Primatope Agreement comprised of upfront consideration of \$10.0 million paid at closing as well as milestone payments of \$5.0 million which had been achieved as of the closing date, and a milestone of \$3.0 million, which we deemed to be probable and estimable and accrued for as of March 31, 2019, each paid or payable in a combination of cash and Class A common shares (inclusive of escrow and holdback amounts) in accordance with the terms and conditions of the Primatope Agreement.

Unallocated research and development expenses were \$10.7 million for the three months ended March 31, 2020 compared to \$12.4 million for the three months ended March 31, 2019. The decrease of \$1.7 million in unallocated research and development expenses was due to a decrease of \$1.9 million related to the termination of our license agreement with Novo Nordisk, or Novo, and the cessation of our development of KPL-045 and a decrease of \$0.7 million in other expenses including operating costs for our laboratory as well as general research costs. The decrease was offset by an increase of \$0.9 million in personnel-related costs primarily related to share-based compensation and contract staff. Personnel-related costs for the three months ended March 31, 2020 and 2019 included share-based compensation of \$1.8 million and \$1.2 million, respectively.

#### *General and Administrative Expenses*

General and administrative expenses were \$8.5 million for the three months ended March 31, 2020 compared to \$8.4 million for the three months ended March 31, 2019. The increase of \$0.1 million was primarily due to increases of \$0.8 million in personnel-related costs partially offset by a decrease of \$0.7 million in professional fees and other expenses. Personnel-related costs for the three months ended March 31, 2020 and 2019 included share-based

compensation of \$2.4 million and \$1.7 million, respectively, or an increase of \$0.7 million. Professional fees decreased primarily due accounting and legal cost associated to the acquisition of Primatope and other professional fees.

#### *Interest Income*

Interest income was \$0.8 million for the three months ended March 31, 2020 compared to \$1.8 million for the three months ended March 31, 2019. The decrease was due to lower invested balances and lower interest rates on U.S. Treasury notes.

#### *Benefit for Income Taxes*

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 February 4, 2019, we completed a follow-on offering of 2,654,984 Class A common shares at a public offering price of \$18.26 per share and concurrent private placement of 2,000,000 Class A1 common shares at an offering price of \$18.26 per share for aggregate gross proceeds of \$85.0 million. In addition, on March 1, 2019, we completed the sale of 161,126 Class A common shares to the underwriters of the follow-on offering following the exercise in part of their over-allotment option to purchase additional shares at a public offering price of \$18.26 per share for gross proceeds of \$2.9 million. The aggregate net proceeds to us from the follow-on offering and concurrent private placement, inclusive of the over-allotment option exercise, was \$83.0 million after deducting underwriting discounts and commissions and other offering costs.

As of March 31, 2020, we had cash, cash equivalents and short-term investments of \$204.2 million.

#### **Cash Flows**

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

	<b>Three Months Ended</b>	
	<b>March 31,</b>	
	<b>2020</b>	<b>2019</b>
	<b>(in thousands)</b>	
Net cash used in operating activities	\$ (31,796)	\$ (63,256)
Net cash provided by (used in) investing activities	88,790	(9,278)
Net cash provided by financing activities	2,414	83,187
Net increase in cash and cash equivalents and restricted cash	<u>\$ 59,408</u>	<u>\$ 10,653</u>

#### *Operating Activities*

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, a \$7.0 decrease in accrued expenses and other liabilities, a \$0.4 million decrease in operating lease liabilities, and a \$0.6 million increase in prepaid expenses and other current assets, partially offset by a \$0.1 million increase in other long-term liabilities. The decrease in accrued expense and other liabilities is primarily due to the cash payment of the 2019 employee bonuses. The increase in prepaid expenses and other current assets was due to increases in prepaid expenses to CROs related to our clinical trials and an increase for the income tax receivable.

During the three months ended March 31, 2019, operating activities used \$63.3 million of cash, primarily resulting from our net loss of \$65.8 million, partially offset by non-cash charges of \$9.6 million and net cash used in our operating assets and liabilities of \$7.0 million. Net cash used in our operating assets and liabilities for the three months ended March 31, 2019 consisted of a \$0.4 million decrease in accrued expenses and a \$3.3 million decrease in accounts payable, a \$0.3 million decrease in operating lease liabilities, a \$1.5 million increase in prepaid expenses and other current assets, partially offset by a \$0.5 million increase in other long-term liabilities. The decrease in accrued expenses was due primarily to a \$2.0 million decrease in accrued milestones, which consisted of milestone payments of \$15.0 million, partially offset by accrued milestones of \$13.0 million. The decrease in accounts payable was primarily due to the timing of vendor invoicing and payments. The increase in prepaid expenses and other current assets was due to interest receivable and prepaid expenses to CMOs related to manufacturing development and CROs related to our clinical trials. The decrease in operating lease liabilities was due to monthly payments for our right of use assets. The increase in other long-term liabilities relates to the portion of consideration to be paid in Class A common shares that was held back in accordance with the terms and conditions of the Primatope Agreement.

#### *Investing Activities*

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.

During the three months ended March 31, 2019 investing activities used \$9.3 million of cash, consisting of \$1.6 million of purchases of property and equipment and \$181.8 million of purchases of short-term investments offset by \$174.1 million from proceeds of maturities of short-term investments.

#### *Financing Activities*

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.

During the three months ended March 31, 2019, net cash provided by financing activities was \$83.2 million, consisting of \$83.0 million from proceeds from our issuance and sale of Class A common shares in a follow-on public offering, inclusive of the concurrent issuance and sale of Class A1 common shares in a private placement, and exercise in part of the underwriters' over-allotment option to purchase additional Class A common shares and concurrent issuance after deduction of underwriting commissions and discounts and other offering costs.

#### **Funding Requirements**

We expect to incur significant expenses in connection with our ongoing activities, particularly as we advance the clinical trials, preclinical activities of our product candidates and prepare for commercial operations. 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 clinical trials for riloncept, mavrilimumab, vixarelimab and KPL-404, as applicable;
- manufacture, or have manufactured on our behalf, our preclinical and clinical drug material and develop processes for late-stage and commercial manufacturing;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- establish a sales, marketing, medical affairs and distribution infrastructure to commercialize any product candidates, including riloncept, for which we may obtain marketing approval and intend to commercialize on our own;

- 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. If we receive regulatory approval for riloncept or our other product candidates, pursue additional indications for our existing product candidates, we expect to incur significant commercialization expenses related to product manufacturing, sales, marketing and distribution, depending on where we choose to commercialize.

Because of the numerous risks and uncertainties associated with research, development and commercialization of biologic 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;
- 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;
- the ability to receive additional non-dilutive funding;
- the revenue, if any, received from commercial sale of our products, should any of our product candidates receive marketing approval;
- the costs of preparing, filing and prosecuting patent applications, maintaining and enforcing our intellectual property rights and defending intellectual property-related claims;
- our ability to establish and maintain 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, 2020, 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 12 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, 2020, 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.

### **Emerging Growth Company Status**

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

### **Off-Balance Sheet Arrangements**

We did not have during the periods presented, and we do not currently have, any off-balance sheet arrangements, as defined in the rules and regulations of the 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, 2020, 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. 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, 2020.

#### ***Changes in Internal Control over Financial Reporting***

There was no change in our internal control over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act) during the three months ended March 31, 2020 that has materially affected, or is 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 audited 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 with a limited operating history and have not generated any revenue from product sales. We have incurred significant operating losses since our inception and anticipate that we will incur continued losses for the foreseeable future.***

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. To date, we have invested substantially all of our efforts and financial resources in identifying, acquiring, in-licensing and developing our product candidates, including commencing and conducting clinical trials and providing general and administrative support for these operations. Our future success is dependent on our ability to develop, obtain regulatory approval for and successfully commercialize one or more of our product candidates. We have not yet demonstrated our ability to successfully conduct and complete a Phase 3 or other pivotal clinical trial, obtain regulatory approvals, manufacture a commercial-scale drug, or conduct sales and marketing activities. We currently generate no revenue from sales of any products, and we may never be able to develop or commercialize a marketable product. Consequently, predictions about our future success or viability could be more accurate if we had a longer operating history.

We have incurred significant losses related to expenses for research and development and our ongoing operations. Our net losses for the three months ended March 31, 2020 and 2019 were \$26.4 million and \$65.8 million, respectively. As of March 31, 2020, we had an accumulated deficit of \$382.5 million. We expect to continue to incur losses for the foreseeable future, and we anticipate these losses will increase substantially as a result of many factors, including:

- our research and preclinical and clinical development of our product candidates, including our global, pivotal Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, named RHAPSODY for which we have completed enrollment, our global Phase 2 clinical trial with mavrilimumab for the treatment of giant cell arteritis, or GCA, for which we have completed enrollment, our potential registrational clinical development program for mavrilimumab for the treatment of the novel coronavirus 2019, or COVID-19, pneumonia and hyperinflammation, our Phase 2a clinical trial with vixarelimab in prurigo nodularis and our exploratory Phase 2 clinical trial for vixarelimab in diseases characterized by chronic pruritus, as well as our Phase 1 clinical trial in healthy volunteers for KPL-404;
- initiating potential additional preclinical studies and clinical trials for our product candidates;
- 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;
- establishing a sales, marketing and distribution infrastructure to commercialize products for which we may obtain marketing approval;
- seeking to identify, assess and study new or expanded indications for our product candidates, new or alternative dosing levels and frequency for our product candidates, or new or alternative administration of our 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;
- making milestone or other payments under any current or future license, 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, our product development and planned future commercialization efforts; and
- experiencing delays or encountering issues with any of the above, including but not limited to 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.

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 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 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 advancing our product candidates through research, preclinical and clinical development, including through our global, pivotal Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, named RHAPSODY, for which we have completed enrollment, our global Phase 2 clinical trial for mavrilimumab for the treatment of GCA for which we have completed enrollment, our potential registrational clinical development program for mavrilimumab for the treatment of COVID-19 pneumonia and hyperinflammation, our Phase 2a clinical trial for vixarelimab for the treatment of prurigo nodularis, our exploratory Phase 2 clinical trial with vixarelimab in diseases characterized by chronic pruritus, and our Phase 1 clinical trial with KPL-404 in healthy volunteers. We expect our expenses to increase in connection with our ongoing activities as we continue the research and development of our product candidates, establish and expand our sales, marketing and distribution capabilities, infrastructure and organization, or enter into agreements with third parties to conduct one or more of these commercialization activities. In addition, if we obtain marketing approval for any of our product candidates, we expect to incur significant additional commercialization expenses related to manufacturing, product sales, marketing and distribution. As our product candidates progress through development and towards commercialization, we will need to make milestone payments and, if successful, eventually make 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. 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 development process for our product candidates is highly uncertain, and we cannot estimate with certainty the actual amounts necessary to successfully complete the development, regulatory approval process and commercialization of our product candidates. Our operating plans may change as a result of many factors currently unknown to us, and we may need to seek additional funds sooner than expected, through public or private 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:

- the results from, and the time and cost necessary for, completing our global, pivotal Phase 3 clinical trial for riloncept in recurrent pericarditis, RHAPSODY, our global Phase 2 clinical trial for mavrilimumab in GCA, our collaborators' investigator-initiated studies in the United States and Italy and our potential clinical development program for mavrilimumab for the treatment of COVID-19 pneumonia and hyperinflammation, the planned Phase 2 clinical trial evaluating the combination of Yescarta (axicabtagene ciloleucel) and mavrilimumab in relapsed or refractory large B-Cell lymphoma under our clinical collaboration with Kite, a Gilead Company, or Kite, for which Kite will be the sponsor and responsible for the conduct of the trial, our Phase 2a clinical trial for vixarelimab in prurigo nodularis, our exploratory Phase 2 clinical trial for vixarelimab in diseases characterized by chronic pruritus, and our Phase 1 clinical trial in healthy volunteers for KPL-404;
- 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 U.S. Food and Drug Administration, or 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 costs and timing of transferring manufacturing technology to third-party manufacturers, producing product candidates to support clinical trials and preparing to manufacture product candidates on a commercial scale, as well as producing riloncept in potential new final form configurations;
- the timing and amount of milestone and other payments we must make under our agreements with Regeneron Pharmaceuticals, Inc., or 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;
- our ability to successfully commercialize any of our product candidates, including the cost and timing of establishing and expanding our sales, marketing and distribution capabilities, infrastructure and organization or entering into agreements with third parties to conduct one or more of these activities for any of our product candidates, if approved or in anticipation of such approval;
- the amount of sales revenues from our product candidates, if approved, including the sales price and the availability of coverage and adequate third-party reimbursement;
- competitive and potentially competitive products and technologies and patients' and physicians' receptivity to our product candidates and the technology underlying them in light of competitive products and technologies;

- 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 cash requirements for seeking to identify, assess and study new or expanded indications for our product candidates, new or alternative dosing levels or frequency for our product candidates, or new or alternative administration of our product candidates, including method, mode or delivery device;
- the time and cost necessary to respond to technological and market developments;
- the costs of filing, prosecuting, defending and enforcing any patent claims and other intellectual property rights;
- any product liability or other lawsuits related to our product candidates or any related activities;
- the costs associated with being a public company;
- 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 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 is adversely impacting the global economy 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, 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 product candidate. We may also be unable to expand our operations or otherwise capitalize on our business opportunities or may be required to relinquish rights to our product candidates or products. Any of these occurrences could materially affect our business, financial condition and results of operations.

***Raising additional capital may cause dilution to our shareholders, restrict our operations or require us to relinquish rights to our technologies or product candidates.***

Until such time as we can generate substantial product revenues, if ever, we expect to finance our cash needs through 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. 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 Product Development**

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

We do not currently generate any revenue from sales of any products, and we may never be able to develop or commercialize marketable products. 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. We have four product candidates 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 on either indirect data primarily collected by other companies or 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 riloncept is approved and marketed for human use for the treatment of cryopyrin-associated periodic syndrome, or CAPS, in the United States by Regeneron, we are studying riloncept for the treatment of a different indication called recurrent pericarditis, which is in a global, pivotal Phase 3 clinical trial, RHAPSODY. Mavrilimumab has been through Phase 2 clinical trials conducted by MedImmune for the treatment of rheumatoid arthritis, or RA, but our global Phase 2 clinical trial with mavrilimumab is for the treatment of GCA. Further, we are engaging with the FDA on our potential registrational clinical development program for mavrilimumab for the treatment of COVID-19 pneumonia and hyperinflammation. Vixarelimab is being studied in a Phase 2a clinical trial in prurigo nodularis and an exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus. In addition, KPL-404 has progressed into a Phase 1 clinical trial in healthy volunteers. 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 have not submitted, and we may never submit marketing applications to the FDA or comparable regulatory authorities outside of the United States for our product candidates. We cannot be certain that any of our product candidates will be successful in clinical trials or receive regulatory approval. Further, our product candidates may not receive regulatory approval even if we complete a successful clinical trial. We may also determine that the potential product and commercial profile of any of our product candidates would not ultimately be commercially successful and could therefore elect not to continue its development. If we do not receive regulatory approvals for one or more of our product candidates, we may not be able to continue our operations.

Each of our product candidates require additional preclinical or clinical development, regulatory approval in one or more jurisdictions, manufacturing capacity and expertise, successful manufacture of clinical supply, building an organization to support commercialization, substantial investment and significant marketing efforts before we will be able to generate any revenue from product sales. The success of our product candidates or potential future product candidates depends upon several factors, including the following:

- 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;
- 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 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;
  - 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 new third-party facilities to support future development activities and commercialization that are operated by contract manufacturing organizations, or CMOs, in a manner compliant with all regulatory requirements;
  - successful manufacture of sufficient supplies of our product candidates within approved specifications for purity and efficacy from our facility and from our CMOs 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;
  - successful commercial launch of our product candidates, if and when approved;
  - 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, we could experience significant delays in, or an inability to, successfully commercialize our product candidates, which would materially harm our business. If we do not receive regulatory approvals for one or more of our product candidates, we may not be able to continue our operations. Even if we successfully obtain regulatory approvals to manufacture and market our product candidates, our revenues will be dependent, in part, upon the size of the markets in the territories for which we gain regulatory approval and have commercial rights. If the markets for patient subsets that we are targeting are smaller than we estimate, we may not generate projected revenue levels from sales of such products, if approved.

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

Before obtaining marketing approval from regulatory authorities for the sale of our product candidates, we must conduct extensive clinical trials to demonstrate the safety and efficacy of our product candidates in humans. Clinical testing is expensive, time consuming and uncertain as to outcome. We are conducting a global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, RHAPSODY, for which we have completed enrollment, a global Phase 2 clinical trial with mavrilimumab for the treatment of GCA for which we have completed enrollment, our potential registrational clinical development program for mavrilimumab for the treatment of COVID-19 pneumonia and hyperinflammation, a Phase 2a clinical trial with vixarelimab in prurigo nodularis, an exploratory Phase 2 clinical trial with vixarelimab in diseases characterized by chronic pruritus, and a Phase 1 clinical trial with KPL-404 in healthy volunteers. We cannot guarantee that any of our current or potential future clinical trials will be conducted as planned or completed on schedule, if at all.

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 may 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 options. 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. 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 demonstrate the safety and effectiveness of mavrilimumab at the 26 weeks of dosing stipulated in our Phase 2 clinical trial, and 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. In addition, the development of mavrilimumab in other potential indications, such as the treatment of COVID-19 pneumonia and hyperinflammation and relapsed or refractory large B-Cell lymphoma in combination with Yescarta (axicabtagene ciloleucel), could increase the possibility of identification of adverse safety results that impact our development of mavrilimumab for 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, or New Drug Applications 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. We do not know whether any of our future clinical trials will begin as planned or any of our current or future clinical trials will be completed on schedule, if at all.

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 may 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 may prevent commencement or successful completion of clinical development of our product candidates as planned, 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 among other things;
- clinical trial sites or patients withdrawing from a clinical trial;

- 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 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 and the business operations of our 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 recent COVID-19 pandemic.

Many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the delay or denial of regulatory approval of our product candidates.

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

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.

Clinical trial delays could also shorten any periods during which our products have patent protection and may allow our competitors to bring products to market before we do, which could impair our ability to obtain orphan exclusivity 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.

***The COVID-19 pandemic, and measures taken in response to the pandemic, could adversely impact our current or planned preclinical studies and clinical trials.***

The COVID-19 pandemic, and measures taken in response to the pandemic, could impact 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 disruptions that could severely 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 global shipping that may affect the transport of clinical trial materials;
- 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 a reprioritizing of the focus of such resources on clinical trials for product candidates with the potential for treatment or prevention of COVID-19-related conditions;
- 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;
- impeding, delaying, limiting or preventing activities required for Regeneron to transfer its BLA for rilonacept to us in the United States for CAPS following positive Phase 3 trial data, if any, to enable us to file a supplemental BLA, or sBLA, with the FDA for rilonacept in the United States for recurrent pericarditis;
- impeding, delaying, limiting or preventing the production, delivery or release of the potential commercial supply of rilonacept, including due to disruptions at Regeneron's manufacturing facilities that produce rilonacept, staffing shortages or reprioritizations, production slowdowns or stoppages or interruptions in global shipping;
- causing interruptions or delays at the FDA, or other regulatory authorities, which could result in delays in review and approval of 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 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. The COVID-19 pandemic continues to rapidly evolve. The extent to which the pandemic impacts our current and planned preclinical studies and clinical trials will depend on future developments, which are highly uncertain and cannot be predicted with confidence, such as the duration of the pandemic, the scope and duration of the measures taken in response to the pandemic, including business and travel restrictions and social distancing, the effectiveness of such measures, and the impact of any easing of such measures.

***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 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, 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 clinical trials may compete with other clinical trials for product candidates that are in the same therapeutic areas as our product candidates, and this competition may further reduce the number and types of patients available to us because some patients who might have opted to enroll in our trials may instead opt to enroll in a trial being conducted by one of our competitors. Since the number of qualified clinical investigators is limited, we may conduct some of our clinical trials at the same clinical trial sites that some of our competitors use, which 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 COVID-19, may delay or prevent patient enrollment. For example, our CRO for the Phase 1 clinical trial of KPL-404 notified us that they temporarily paused enrollment and dosing of new patients, but we do not expect this temporary pause will impact our timeline to report data from the trial in the second half of 2020. However, if the CRO does not restart the enrollment and dosing process for new patients when expected or at all, it could impact the timeline of the clinical trial and the breadth of data we may be able to generate.

Accordingly if we encounter these or other difficulties in enrollment we may experience delays or 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;
- 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 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. 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 and the

timeline for recruiting patients, conducting trials and obtaining regulatory approval of our product candidates may be 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 product candidates may cause undesirable side effects or have other safety risks that could delay or prevent their regulatory approval, limit the commercial profile of an approved label or result in significant negative consequences, including withdrawal of approval, following any potential marketing approval.***

Treatment with our product candidates may produce undesirable side effects or adverse reactions or events. Undesirable side effects caused by our product candidates could cause us or regulatory authorities to interrupt, delay or halt clinical trials and could result in 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 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 example, some common side effects of riloncept include, cold symptoms, nausea, stomach pain, diarrhea, numbness or tingly feeling and injection-site reaction. IL-1 blockade may interfere with immune response to or delay symptomatology and diagnosis of infections. Serious, life-threatening infections have been reported in patients taking riloncept. In our open-label Phase 2 proof-of-concept clinical trial of riloncept for recurrent pericarditis, the most common AEs were gastrointestinal disorders and injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment.

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 product candidates for any or all targeted indications.

In addition, subsequent to MedImmune's original IND submission for RA and the availability of additional clinical safety data that MedImmune generated in human clinical trials conducted outside of the United States for RA, the FDA 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 options. Further, 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. 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 demonstrate the safety and effectiveness of mavrilimumab at the 26 weeks of dosing stipulated in our Phase 2 clinical trial, and 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 our Phase 2a clinical trial of vixarelimab in in prurigo nodularis, vixarelimab was well-tolerated by all subjects and no dose-limiting adverse experiences were observed. There were no serious adverse events or atopic dermatitis flares. However, in our repeated-single-dose Phase 1b clinical trial of vixarelimab, there were no serious AEs. However, there were more atopic dermatitis flares in the vixarelimab-treated population versus placebo (47.6% versus 4.5%) through the 12-week treatment period; all subjects who experienced a flare were successfully managed with topical corticosteroids. Vixarelimab was otherwise well-tolerated by all subjects.

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 and in relapsed or refractory large B-Cell lymphoma in combination with Yescarta (axicabtagene ciloleucel), could increase the possibility of identification of adverse safety results that impact our development of mavrilimumab for GCA or any other indication.

Additionally, clinical trials by their nature utilize a sample of the potential patient population. Certain rare and severe side effects associated with our product candidates may only be uncovered with a significantly larger number of patients exposed to the product candidates. If one or more of our product candidates receives 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 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 the companies from whom we have licensed or acquired, or may in the future license or acquire, our product candidates in our later clinical trials, we may be unable to successfully develop, obtain regulatory approval for and commercialize our product candidates.***

Positive results from our preclinical studies and any positive results we may obtain from our early clinical trials of our product candidates, or from the clinical trials conducted by the companies from whom we 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 clinical trials of

our product candidates may not be replicated in our subsequent preclinical studies or clinical trial results. The mechanisms of action of our product candidates may not prove to be safe or effective to treat the diseases that we are studying. Further, the safety and efficacy of our product candidates have not been established for the indications in which we are developing them, and we cannot provide any assurance that their development will be successful. For example, although rilonacept is FDA approved for the treatment of CAPS, and mavrilimumab has been studied in Phase 2 clinical trials for the treatment of RA, their safety and efficacy have not been determined in the indications we are pursuing, recurrent pericarditis or GCA, respectively, and each may fail to receive regulatory approval for those indications.

Many companies in the pharmaceutical and biotechnology industries have suffered significant setbacks in 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 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.

***Preliminary, interim 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 and are subject to audit and verification procedures that could result in material changes in the final data.***

From time to time, we may publicly disclose interim, top-line or preliminary data from our clinical trials, which are based on a preliminary analysis of then-available data and are subject to change following a more comprehensive review of the data related to the particular study or trial. We also make assumptions, estimations, calculations and conclusions as part of our analyses of data, and we may not have received or had the opportunity to fully and carefully evaluate all data. As a result, the top-line or preliminary results that we report may differ from future results of the same studies, or different conclusions or considerations may qualify such results, once additional data have been received and fully evaluated. Top-line or preliminary data also remain subject to audit and verification procedures that may result in the final data being materially different from the top-line or preliminary data we previously published. As a result, top-line and preliminary data should be viewed with caution until the final data are available.

From time to time, we may also disclose interim data from our preclinical studies and clinical trials. Interim data from clinical trials that we may complete are subject to the risk that one or more of the clinical outcomes may materially change as patient enrollment continues and more patient data become available. Adverse differences between interim data and final data could significantly harm our business prospects.

Further, others, 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 or product and our company in general. In addition, the information we choose to publicly disclose regarding a particular study or clinical trial is based on what is typically extensive information, and our shareholders or others may not agree with what we determine is material or otherwise appropriate information to include in our disclosure.

If the interim, top-line or preliminary data that we report differ from actual results, or if others, 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, operating results, prospects or financial condition. Further, disclosure of preliminary, interim or top-line data by us or differences between preliminary, interim or top-line data and 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 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 product candidates and our ability to generate revenue will be materially impaired.***

Our product candidates and the activities associated with their development and commercialization, including their trial design, testing, manufacture, safety, efficacy, recordkeeping, 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 product candidates, we must obtain marketing approval. We have not received approval or clearance to market any of our product candidates from regulatory authorities in any jurisdiction and it is possible that none of our product candidates or any product candidates we may seek to develop in the future will ever obtain regulatory approval or clearance. We have only limited experience in filing and supporting the applications necessary to gain regulatory approvals and may need to rely on third-party CROs and regulatory consultants to assist us in this process. Securing regulatory approval requires the submission of extensive 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 authority. Our product candidates may not be effective, may be only moderately effective or may prove to have undesirable or unintended side effects, toxicities or other characteristics that may preclude our obtaining marketing approval or prevent or limit commercial use. The FDA and other regulatory authorities have substantial discretion in the approval process, and determining when or whether regulatory approval will be obtained for a product candidate. Even if we believe the data collected from clinical trials are promising, such data may not be sufficient to support approval by the FDA or any other regulatory authority. 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, clinical or other trials.

In addition, we plan to have Regeneron transfer its BLA for riloncept in the United States for the treatment of CAPS following positive clinical data readout, if any, from our global, pivotal Phase 3 clinical trial, RHAPSODY, to enable us to file an sBLA with the FDA to seek approval for the commercial marketing of riloncept in the United States for the treatment of recurrent pericarditis. The rights with respect to CAPS would remain with Regeneron until the date that we receive such approval from the FDA, if at all. In order for the BLA to be transferred to us, we must coordinate numerous activities with Regeneron in advance and take over certain responsibilities and obligations with respect to owning the BLA. The process to transfer a BLA is complex and resource intensive, and may be more so than we anticipate. Further, if Regeneron or we do not perform in accordance with our expectations, or we are not ready to assume the responsibilities on a timely basis, or the timing of the BLA transfer is otherwise delayed, our ability to file an sBLA with the FDA may be delayed, which could delay FDA approval or our commercialization of riloncept, and our ability to generate revenue from riloncept could be materially impaired.

Approval by the FDA in the United States, if obtained, does not ensure approval by regulatory authorities in other countries or jurisdictions. Our product candidates could be delayed in receiving, or fail to receive, regulatory approval 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;
- 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 study 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 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 collect additional data or conduct additional clinical studies, for example, based on FDA feedback, we anticipate that to help inform the risk-benefit profile for the use of mavrilimumab in GCA, we will need to demonstrate the effectiveness and safety of mavrilimumab at the 26 weeks of dosing stipulated in our global Phase 2 clinical trial, and 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;
- the FDA or comparable regulatory authorities in other jurisdictions could require us to conduct additional clinical studies 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 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 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 product candidates, regulatory authorities may approve any of our product candidates for fewer or more limited indications than we request. For example, in connection with our 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 product candidates.

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

***Our 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, rilonacept was approved as a biological product under a BLA for the treatment of CAPS in 2008, and we believe it should qualify for the 12-year period of exclusivity against any biosimilars. However, there is a risk that this exclusivity could be shortened due to Congressional action or otherwise, or that the FDA will not consider rilonacept, or any of our other product candidates, to be reference products for competing products, potentially creating the opportunity for generic competition sooner than anticipated. In addition, if our clinical trials are successful, we plan

to submit a sBLA for rilonacept for the treatment of recurrent pericarditis, and the 12-year exclusivity period does not attach to the approval of a sBLA, potentially creating the opportunity for biosimilar competition.

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

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

In order to market any products in a country or territory, we must establish and comply with numerous and varying regulatory requirements of such country or territory regarding safety and efficacy. 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 product candidates in those countries. Satisfying these and other regulatory requirements is costly, time consuming, uncertain and subject to unanticipated delays. In addition, our failure to obtain regulatory approval in any country may delay or have negative effects on the process for regulatory approval in other countries.

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

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

In the 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 may pursue orphan drug designation for certain of our product candidates, we may never receive such designation. Even if we do receive such designation, there is no guarantee that we will enjoy the benefits of such designation.

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

Conducting pivotal clinical trials and preparing, and obtaining marketing approval for, a product candidate is a complicated process. Although members of our management team have participated in pivotal trials and obtained marketing approvals for product candidates in the past while employed at other companies, we as a company have not done so. As a result, such activities may require more time and cost more than we anticipate. Failure to successfully complete, or delays in, our global, pivotal Phase 3 clinical trial in rilonacept or 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 product candidates. In addition, it is possible that the FDA or other government agencies 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 that they are insufficient to obtain marketing approval or clearance of our product candidates. If the FDA or other government agencies do not accept our applications or issue marketing authorizations for our product candidates, they 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 government agencies to approve or grant marketing authorizations. Any delay in obtaining, or an inability to obtain, marketing approvals would delay or prevent us from commercializing our product candidates, generating revenues and achieving and sustaining profitability. If any of these outcomes occur, we may be forced to modify or cease our development efforts for 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, or approved or commercialized in a timely manner or at all, which could negatively impact our business.***

The ability of the FDA or other government agencies 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 ability to hire and retain key personnel and accept the payment of user fees, and other events that may otherwise affect the ability to perform routine functions. Average review times at the agency 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 government agencies may also slow the time necessary for new drugs and biologics or modifications to cleared or approved drugs and 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 foreign inspections of manufacturing facilities and products for a period of time, and subsequently, on March 18, 2020, the FDA announced its intention to temporarily postpone routine surveillance inspections of domestic manufacturing facilities. Government agencies 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 government agencies from conducting their regular inspections, reviews, or other regulatory activities, it could significantly impact the ability of the FDA or other government agencies 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 product candidates and for certain research and other preclinical and clinical development and expect to continue to do so for our commercial supply. This reliance on third parties increases the risk that we may not have sufficient quantities of our product candidates or such quantities at an acceptable cost, which could delay, prevent or impair our development or commercialization efforts.***

We do not currently own or operate any late-stage manufacturing facilities. Although we have built a development and manufacturing facility to produce drug substance to support certain research and other preclinical and early 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 potential commercial manufacture of our product candidates, if approved, as well as label and packaging activities. We rely on these third parties to develop the processes necessary to produce our product candidates at sufficient quality and quantity to support our development and commercialization efforts. Our reliance increases the risk that we will have insufficient quantities of our product candidates or that our product candidates are not produced at an acceptable cost or quality, which could delay, prevent or impair our development or commercialization efforts.

For example, we have a contract with Regeneron to produce riloncept on an exclusive basis for a period of time. Although Regeneron has produced riloncept for commercial use for over ten years, the FDA or other applicable regulatory authorities in other jurisdictions might reevaluate riloncept's current manufacturing process or route of administration in connection with evaluating whether to approve riloncept for a new indication, such as recurrent pericarditis. 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, which could delay, prevent or impair our development or commercialization efforts and increase costs.

If we make manufacturing or formulation changes to our product candidates or change manufacturers or manufacturing processes, we may be unsuccessful in producing products 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. For example, while we have transferred the technology from Medimmune needed for us to manufacture mavrilimumab in our facility, the process needs to be re-developed. There is no guarantee that the new process will produce a comparable product. In addition, as we plan to produce clinical trial material at a CMO, the CMO may be required to adopt different manufacturing protocols or processes. The CMO will need to produce mavrilimumab in a different manufacturing site and at a different scale than ours. We cannot provide any assurance that the technology transfer from Medimmune to us was successful or that our process development or that of the CMO will be successful in producing mavrilimumab in sufficient quantities or of acceptable quality, if at all, which could delay, prevent or impair the development of mavrilimumab.

The facilities used by our CMOs to manufacture our product candidates may be inspected by the FDA and other comparable regulatory authorities in connection with the submission of our marketing applications to, and review by, the FDA or other comparable 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 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 may review the compliance history and performance of our CMOs, we have no control over the ability of our CMOs to maintain adequate quality control, quality assurance and qualified personnel. 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 our product candidates, if approved.

Further, our failure, or the failure of our third-party manufacturers, to comply with applicable regulations could result in sanctions being imposed on us, including clinical holds, fines, injunctions, civil penalties, delays, suspension or withdrawal of approvals, license revocation, seizures or recalls of product candidates, if approved, operating restrictions and criminal prosecutions, any of which could significantly and adversely affect our business and supplies of our product candidates.

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

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

Our product candidates may compete with other product candidates and approved products for access to manufacturing facilities. There are a limited number of manufacturers that operate under cGMP regulations and that might be capable of manufacturing for us. 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. Such projections involve risks and uncertainties and may result in additional costs or delays in manufacturing clinical materials for our product candidates when and if we actually need them.

Any performance failure on the part of our existing or future manufacturers could delay clinical development or marketing approval. If our current CMOs cannot perform as agreed, we may be required to replace such manufacturers. Although we believe that there are several potential alternative manufacturers who could manufacture our product candidates, we may incur added costs and delays in identifying and qualifying any such replacement. Further, Regeneron has an exclusive right to produce rilonacept for a period of time, which could impact our ability to find a replacement manufacturer for rilonacept in a short-period of time if needed.

Finding new CMOs or third-party suppliers involves additional cost and requires our management’s time and focus. In addition, there is typically a transition period when a new CMO commences work. Any significant delay in the supply of our product candidates or the raw materials needed to produce our product candidates, could considerably delay conducting our clinical trials and potential regulatory approval of our product candidates.

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

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

Our research and development activities and our third-party manufacturers' and suppliers' activities involve the controlled storage, use and disposal of hazardous materials owned by us, including the components of our product 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 could cause product shortages, disrupt or delay our clinical trials or regulatory approvals, delay or stop commercialization of our product candidates, and adversely affect our business.***

The manufacture of our product candidates is highly regulated, complex and difficult, requiring a multi-step and controlled process, and even minor problems or deviations could result in 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, KPL-404 and no experience overseeing the manufacturing process of rilonacept. 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 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, which may lead to lawsuits or could delay the introduction of our product candidates to the market.

The manufacture of our product candidates is at high risk of product loss due to contamination, equipment malfunctions, human error or raw material shortages. Deviations from established manufacturing processes could result in reduced production yields, failed batches and other supply disruptions. If microbial, viral or other contaminations are discovered in our product candidates or manufacturing facilities, any related production lot could be lost and the relevant manufacturing facilities may need to close for an extended period of time to investigate and remediate the contaminant.

Many additional factors could cause production interruptions at our facilities or at the facilities of our third-party providers, as well as disruptions in travel or delivery capabilities into and within the countries in which we or our manufacturers produce 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 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 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.

In addition, we plan to have Regeneron transfer its BLA for riloncept in the United States for the treatment of CAPS following positive clinical data readout, if any, from our global, pivotal Phase 3 clinical trial, RHAPSODY to enable us to file an sBLA with the FDA to seek approval for the commercial marketing of riloncept in the United States for the treatment of recurrent pericarditis. The rights with respect to CAPS would remain with Regeneron until the date that we receive such approval from the FDA, if at all. In order for the BLA to be transferred to us, we must coordinate numerous activities with Regeneron in advance and take over certain responsibilities and obligations with respect to owning the BLA. The process to transfer a BLA is complex and resource intensive, and may be more so than we anticipate. Further, if Regeneron or we do not perform in accordance with our expectations, or we are not ready to assume the responsibilities on a timely basis, or the timing of the BLA transfer is otherwise delayed, our ability to file an sBLA with the FDA may be delayed, which could delay FDA approval or our commercialization of riloncept, and our ability to generate revenue from riloncept could be materially impaired.

Any adverse developments affecting the operations of our third-party providers, such as any impact due to the COVID-19 pandemic, could result in a product shortage of clinical or commercial requirements, withdrawal of our product candidates or any approved products, shipment delays, lot failures or recalls. We may also have to write off inventory and incur other charges and expenses for products that fail to meet specifications, undertake costly remediation efforts or seek more costly manufacturing alternatives. Such manufacturing issues could increase our cost of goods, cause us to lose potential revenue, reduce our potential profitability or damage our reputation.

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

The drug substance and drug product used in riloncept, mavrilimumab and vixarelimab are supplied to us from single-source suppliers. For example, Regeneron has a contractual right to be our sole source manufacturer of riloncept 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 riloncept, 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 one or more of our third-party manufacturers' or suppliers' relevant operations, such as due the impact of the COVID-19 pandemic, the supply of the related product candidate will be delayed until such manufacturer or supplier restores the affected facilities or we or they procure alternative manufacturing facilities or sources of supply. Our ability to progress our preclinical and clinical programs could be

materially and adversely impacted if any of the third-party suppliers upon which we rely for preclinical and clinical stage product candidate 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 third-party manufacturers' or suppliers' facilities or equipment may significantly impair our ability to manufacture our product candidates on a timely basis.

Establishing additional or replacement suppliers for the drug substance and drug product used in 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 are able to find a replacement supplier, such replacement supplier would need to be qualified and may require additional regulatory approval, which could result in further delay. While we seek to maintain adequate inventory of the drug substance and drug product used in 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 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 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 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 product candidates could adversely impact or disrupt manufacturing, which would impair our ability to generate revenues from the sale of such product candidates, if approved.

***We rely, and expect to continue to rely, on third parties, including independent investigators and CROs, to conduct our research, 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 conduct or otherwise support our GLP-compliant preclinical studies and GCP-compliant clinical trials for our product candidates properly and on time. We also rely on third parties to conduct other research related to our product candidates. We expect to rely heavily on these parties for execution of clinical trials for our product candidates. While we have agreements governing their activities, we control only certain aspects of these parties' activities and have limited influence over their actual performance. The third parties with whom we contract for execution of our GLP-compliant preclinical studies and our GCP-compliant 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 GLP-compliant preclinical studies and GCP-compliant clinical trials, 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 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 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 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 “The COVID-19 pandemic could adversely impact our preclinical studies and clinical trials.”);
- 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 conduct 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 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, material transfer agreements, collaborative research agreements, consulting agreements or other similar agreements with our collaborators, advisors, employees and consultants prior to beginning research or disclosing proprietary information. These agreements typically limit the rights of the third parties to use or disclose our confidential information, such as trade secrets. Despite the contractual provisions employed when working with third parties, the need to share trade secrets and other confidential information increases the risk that such trade secrets become known by our competitors, are inadvertently incorporated into the technology of others, or are disclosed or used in violation of these agreements. Given that our proprietary position is based, in part, on our know-how and trade secrets, a competitor's discovery of our trade secrets or other unauthorized use or disclosure would impair our competitive position and may harm our business. To the extent that we share trade secrets of third parties that are licensed to us, unauthorized use or disclosure could expose us to liability.

#### **Risks Related to Competition, 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-1 $\alpha$  and IL-1 $\beta$ , anakinra (KINERET), marketed by Swedish Orphan Biovitrum AB, and one product that modulates the signaling of IL-1 $\beta$ , canakinumab (ILARIS), marketed by Novartis Pharmaceuticals Corporation. There is also another therapy which modulates IL-1 $\alpha$  in clinical development for diseases other than recurrent pericarditis from Johnson & Johnson. We expect mavrilimumab, if approved, 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. 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. and UCB, Inc., C.H. Boehringer Sohn AG & Ko. KG and Abbvie Inc., Annelixis Therapeutics LLC, ImmuNext Inc. and Sanofi S.A., Viela Bio 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.

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.***

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. The objective of the Phase 2 trial is to determine the effect of mavrilimumab on the safety of Yescarta. Kite will be the sponsor of this study and responsible for its conduct. We may seek to enter into additional 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 not be favorable to us. In addition, our right to grant a sublicense of intellectual property licensed to us under certain of our current agreements requires the consent of the applicable licensor.

Any 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. 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 to attract, retain and motivate qualified personnel.***

We have a limited operating history and are highly dependent on the research and development, clinical, commercial and business development expertise of members of our executive and senior management teams, as well as the other principal 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 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 or other key employees could impede the achievement of our research, development and commercialization objectives and seriously harm our ability to successfully implement our business strategy. Furthermore, replacing executive officers and key employees may be difficult and may take an extended period of time because of the limited number of individuals in our industry with the breadth of skills and experience required to successfully develop, gain regulatory approval of and commercialize products. Chris Heberlig, our Chief Financial Officer, departed the Company in March 2020 and while we commenced a search for a new Chief Financial Officer, we may experience difficulties or delays in identifying a qualified replacement. These or other changes in our senior management may be disruptive to our business, and, if we are unable to manage an orderly transition, 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 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, expand our facilities and continue to recruit and train qualified personnel. Also, our management may need to divert a disproportionate amount of its attention away from its day-to-day activities and devote a substantial amount of time to managing these development and expansion activities. Due to our limited resources, certain employees may need to perform activities that are beyond their regular scope of work, and we may not be able to effectively manage the development of our company, expansion of our operations or recruit and train qualified personnel. This may result in weaknesses of our infrastructure, give rise to operational mistakes, loss of business opportunities, loss of employees and reduced productivity among remaining employees. The 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 management is unable to effectively manage our expected development and expansion, our expenses may increase more than expected, our ability to generate revenue could be reduced and we may not be able to implement our business strategy. Our future financial performance and our ability to commercialize our product candidates, if approved, and 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 rilonacept, 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 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 rilonacept, an exclusive license under a license agreement with MedImmune, or the MedImmune Agreement, to patent applications and patents relating to mavrilimumab, and an exclusive license under our license agreement with 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 rilonacept,

mavrilimumab, vixarelimab 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 to apply prior to expiration of relevant patents. For example, no patent term extension was obtained in the United States following the FDA's approval of rilonacept for the treatment of CAPS, and the deadline for applying for such extension has passed. Accordingly, patent term extension in the United States based on the FDA's approval of rilonacept for CAPS, or any other indication for which the FDA may grant approval in the future, is unavailable. Further, while patent term extension was awarded for relevant patents in certain European countries following the EMA's approval of rilonacept 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 rilonacept 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 covering rilonacept as a composition of matter have a term that expired in 2019 in the United States, not including patent term adjustment (an adjustment to the term of the U.S. patent to compensate the patentee for delays caused by the USPTO during the examination process), and in 2023 in Europe, 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 or extensions, and in 2027 in Europe, not including any patent term extensions. As a result, our owned and in-licensed patent portfolio may not provide us with adequate and continuing patent protection sufficient to exclude others from commercializing products similar or identical to our product candidates. In such cases, we expect to rely on regulatory exclusivity for our product candidates, such as orphan drug exclusivity, which generally grants seven years of marketing exclusivity under the Federal Food, Drug, and Cosmetic Act, and up to ten years of marketing exclusivity in Europe. While, we may pursue orphan drug designation for our product candidates in the United States, we may not be successful in obtaining such designation or we may not be able to maintain the benefits of the designation. Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent 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 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 our product candidates, riloncept, 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 riloncept. 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 rilonacept in its retained fields of local administration to the eye and ear, oncology, deficiency of the IL-1 receptor, and CAPS. Regeneron may also develop rilonacept in fields to which we have licensed the rights, but we retain the commercial benefit related to that development upon approval of rilonacept in any field that we have licensed. We and Regeneron communicate with each other concerning our related development activities, and we have approval rights over Regeneron's development in the fields that we have licensed, including pericarditis. Outside of the United States and Japan, Regeneron has granted a third-party licensee the right to develop and commercialize rilonacept in CAPS and certain periodic fever syndromes. Regeneron is also developing rilonacept for the treatment of Deficiency of the Interleukin-1 Receptor Antagonist. The development of rilonacept in other fields could increase the possibility of identification of adverse safety results that impact our development of rilonacept for

recurrent pericarditis. In addition, if approved, commercialization of rilonacept in other fields could result in an increased threat of off-label use to compete with the sale of rilonacept to treat these indications, which may diminish sales of rilonacept in fields licensed exclusively to us.

Certain of our agreements may limit or delay our ability to consummate certain transactions, may impact the value of those transactions, or may limit our ability to pursue certain activities. For example, under the MedImmune Agreement, we cannot sublicense the rights licensed or sublicensed to us without the consent of MedImmune and certain applicable third-party licensors, if required by agreements between MedImmune and such third-party licensors. Under 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 agreement.

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

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

Given the vast number of patents in our field of technology, we cannot be certain that we do not infringe existing patents or that we will not infringe patents that may be granted in the future. Many companies have filed, and continue to file, patent applications related to immunomodulation. Some of these patent applications have already been allowed or issued, and others may issue in the future. For example, we are aware of third-party patents that contain claims potentially relevant to certain therapeutic uses of mavrilimumab and 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 and consultants, and invention assignment agreements with our consultants, scientific advisors and employees, we may not be able to prevent the unauthorized disclosure or use of our technical know-how or other trade secrets by the parties to these agreements. Moreover, we cannot guarantee that we have entered into such agreements with each party that may have or have had access to our confidential information or proprietary technology and processes. Monitoring unauthorized uses and disclosures is difficult, and we do not know whether the steps we have taken to protect our proprietary technologies will be effective. If any of the collaborators, scientific advisors, employees, contractors and consultants who are parties to these agreements breaches or violates the terms of any of these agreements, we may not have adequate remedies for any such breach or violation, and we could lose our trade secrets as a result. Moreover, if confidential information that is licensed or disclosed to us by our partners, collaborators, or others is inadvertently disclosed or subject to a breach or violation, we may be exposed to liability to the owner of that confidential information. Enforcing a claim that a third party illegally obtained and is using our trade secrets, like patent litigation, is expensive and time consuming, and the outcome is unpredictable. In addition, courts outside the United States are sometimes less willing to protect trade secrets.

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

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

Our trade secrets could otherwise become known or be independently discovered by our competitors. Competitors could purchase our product candidates and attempt to replicate some or all of the competitive advantages

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

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

Our registered or unregistered trademarks or trade names may be challenged, infringed, circumvented or declared generic or determined to be infringing on other marks. We may not be able to protect our rights to these trademarks and trade names, which we need to build name recognition among potential partners or customers in our markets of interest. At times, competitors or other third parties may adopt trade names or trademarks similar to ours, thereby impeding our ability to build brand identity and possibly leading to market confusion. In addition, there could be potential trade name or trademark infringement claims brought by owners of other registered trademarks or trademarks that incorporate variations of our registered or unregistered trademarks or trade names. Over the long term, if we are unable to establish name recognition based on our trademarks and trade names, then we may not be able to compete effectively and our business may be adversely affected. Our efforts to enforce or protect our proprietary rights related to trademarks, trade secrets, domain names, copyrights or other intellectual property may be ineffective and could result in substantial costs and diversion of resources and could adversely affect our business, financial condition, results of operations and prospects.

***We have not yet registered trademarks for a commercial trade name for our 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.

#### **Risks Related to Commercialization**

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

The precise incidence and prevalence for all the conditions we aim to address with our programs are not known with specificity. Our projections of both the number of people who have these diseases, as well as the subset of people with these diseases who have the potential to benefit from treatment with our product candidates, are based on our extrapolation from available population data and estimates. These estimates have been derived from a variety of sources, including the scientific literature, surveys of clinics, patient foundations, pharmacy claims analyses, 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. As a result, the number of patients who may benefit from our product candidates may turn out to be lower than expected.

The total addressable market for any of our product candidates will ultimately depend upon, among other things, the diagnostic criteria and applicable patient population included in the final label for the 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.

***If we are unable to establish and expand our sales, marketing and distribution capabilities, either directly or through agreements with third parties, we may not be successful in commercializing our product candidates, if approved, thus potentially impairing commercial potential for our product candidates to generate any revenue.***

We have never sold, marketed or distributed any therapeutic products. To achieve commercial success for any approved product candidate, we must build our sales, marketing, distribution, managerial and other non-technical capabilities or make arrangements with third parties to perform these services. For example, we are currently undertaking plans to establish and develop our sales, marketing, and distribution capabilities and infrastructure to directly commercialize rilonacept, in anticipation of approval.

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 product launch. If the commercial launch of a product candidate for which we recruit a sales force and establish marketing capabilities is delayed or does not occur for any reason, we would have prematurely or unnecessarily incurred these commercialization expenses. This may be costly, and our investment would be lost if we are unable to retain or reposition our sales and marketing personnel.

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

- our inability to recruit and retain adequate numbers of effective sales, marketing and access personnel;
- the inability of sales personnel to obtain access to physicians and for an adequate number of physicians to prescribe 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 equip medical and sales personnel with effective materials, including medical and sales literature to help them educate physicians and other healthcare providers regarding applicable diseases relevant to our future products;
- our inability to effectively distribute products in a timely manner;
- our inability to develop or obtain sufficient operational functions and infrastructure to support our commercial activities; and
- unforeseen costs and expenses associated with creating a sales, marketing, and access organization.

If we enter into arrangements with third parties to perform sales, marketing, distribution and other commercial support services, our product revenues or the profitability of these revenues to us are likely to be lower than if we were

to market and sell any product candidates that we develop ourselves. In addition, we may not be successful in entering into arrangements with third parties to sell and market our product candidates or may be unable to do so on terms that are favorable to us. We may have little contractual control over such third parties, and any of them may fail to devote the necessary resources and attention to sell and market our product candidates effectively. Furthermore, developing a sales, marketing and access organization requires significant investment, is time consuming and could delay the launch of our product candidates. We may not be able to build an effective sales, marketing and access organization in the United States, the EU or other key markets. If we do not establish sales, marketing and access capabilities successfully, either on our own or through arrangements with third parties, we will not be successful in commercializing our product candidates. Further, our business, results of operations, financial condition and prospects will be materially adversely affected.

***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 our product candidates and result in lower than anticipated future revenue.***

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. This has resulted in limitations on certain aspects of our commercialization strategy, including interacting with third-party payors, physicians and patient advocacy groups to build disease awareness at industry conferences or otherwise, convening advisory boards and other research activities as well as recruiting qualified candidates to enhance our commercial operations, which, if prolonged, may impede the effective commercialization of our product candidates and result in lower than anticipated future revenue.

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

- the timing of market introduction;
- the number and clinical profile of competing products;
- the potential and perceived advantages or disadvantages of our product candidates relative to alternative treatments;
- the clinical indications for which our product candidates are approved;
- our ability to provide acceptable evidence of safety and efficacy;
- the prevalence and severity of any side effects;
- product labeling or product insert requirements of the FDA or other regulatory authorities;
- limitations or warnings contained in the labeling approved by the FDA or other regulatory authorities;
- 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
- other potential advantages over alternative treatment methods.

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

***The successful commercialization of our product candidates will depend in part on the extent to which 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 product candidates, if approved, could limit our ability to market those products and decrease our ability to generate revenue.***

Our ability to commercialize any of our product candidates successfully, particularly in orphan or rare disease indications, will depend in part on the availability of favorable coverage and the adequacy of reimbursement for the product candidate and alternative treatments from third-party payors (e.g., governmental authorities, private health insurers and other organizations). Obtaining coverage and adequate reimbursement is contingent on our ability to:

- obtain 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. To commercialize our product candidates successfully, we will be required to have sufficient expertise and resources to execute on the respective product candidate's coverage and reimbursement strategy, which we cannot be certain we will be able to do.

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 patient-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 more limited than the purposes for which the product 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, any product candidate for which we obtain marketing approval. 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 any product candidate 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. It is possible that third-party payors will change the clinical comparators that serve as benchmarks for determining relative value. 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 support a value/benefit assessment and refuse to provide coverage and reimbursement, thereby impacting or preventing the progression to a price negotiation. 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 our product candidates successfully.

Third-party payors are also introducing more challenging price negotiation tactics, including in re-visiting established coverage and reimbursement in cases 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 offer to cover the cost of the alternative product. Even if we show improved efficacy or improved convenience of administration with our product candidates, pricing of competitive products may limit the amount we will be able to charge for our product candidates. 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. In other cases, 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 tactics could have a negative impact on our ability to commercialize our product candidates successfully.

***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 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, 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 cuts 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 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 our product candidates and adequate reimbursement.

***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. For example, we are conducting a global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis. Although we do not have immediate plans to pursue the commercialization of rilonacept for recurrent pericarditis outside of the United States, we are evaluating the opportunities for the development and commercialization of our product candidates in 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 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 are relying 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.

***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 on January 31, 2020 and entered into a transition period during which it will continue its ongoing and complex negotiations with the European Union relating to the future trading relationship between the parties. Significant political and economic uncertainty remains about whether the terms of the relationship will differ materially from the terms before withdrawal, as well as about the possibility that a so-called "no deal" separation will occur if negotiations are not completed by the end of the transition period.

These developments, or the perception that any of them could occur, have had and may continue to have a material adverse effect on global economic conditions and the stability of global 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. For example, there may be uncertainty regarding how clinical trials may be regulated. This could have a material adverse effect on our business, financial condition and results of operations and reduce the price of our shares.

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

If the FDA or a comparable regulatory authorities outside of the United States approves any of our product candidates, we will be 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 approved product candidates, including both federal and state requirements in the United States and requirements of comparable regulatory authorities outside of the United States.

Manufacturers and manufacturers' facilities are required to comply with extensive FDA, and comparable regulatory authorities outside of the United States, requirements, including ensuring that quality control and manufacturing procedures conform to cGMP regulations. As such, we and our CMOs will be subject to user fees and continual review and inspections to assess compliance with cGMP and adherence to commitments made in any BLA or MAA. Accordingly, we and 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, if the FDA or a comparable regulatory authority

outside of the United States approves any of our product candidates we will have to 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 or co-marketers fails to comply with regulatory requirements, the regulatory authorities could take various actions. These include imposing fines on us, imposing restrictions on our product or its manufacture and requiring us to recall or remove 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 current U.S. presidential administration has taken several executive actions, including the issuance of a number of Executive Orders, that could impose significant burdens on, or otherwise materially delay, the FDA's ability to engage in routine oversight activities such as implementing statutes through rulemaking, issuance of guidance and review and approval of marketing applications. It is difficult to predict how these Executive Orders will be implemented, and the extent to which they will impact the FDA's ability to exercise its regulatory authority. If these executive actions impose restrictions on the FDA's ability to engage in oversight and implementation activities in the normal course, our business may be negatively impacted. In addition, if we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we are not able to maintain regulatory compliance, we may lose any marketing approval that we may have obtained and we may not achieve or sustain profitability.

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

Although we do not currently have any products on the market, as we near commercialization and as we begin commercializing our product candidates, if approved, we will be 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 providers, physicians and third-party payors will play a primary role in the recommendation and prescription of any product candidates for which we obtain marketing approval. Our future arrangements with third-party payors and customers may expose us to broadly applicable fraud and abuse and other healthcare laws and regulations that may constrain the business or financial arrangements and relationships through which we market, sell and distribute our product candidates for which we obtain marketing approval.

Restrictions under applicable federal and state healthcare laws and regulations, include the following:

- the U.S. federal Anti-Kickback Statute, which prohibits, among other things, persons from knowingly and willfully soliciting, offering, receiving or providing remuneration, directly or indirectly, in cash or in kind, to induce or reward either the referral of an individual for, or the purchase, order or recommendation of, any good or service, for which payment may be made under federal and state healthcare programs such as Medicare and Medicaid. A person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation. The U.S. federal Anti-Kickback Statute has been interpreted to apply to arrangements between pharmaceutical manufacturers on the one hand and prescribers, purchasers and formulary managers on the other hand;
- the U.S. federal False Claims Act and civil monetary penalties laws, which, among other things, impose criminal and civil penalties, including through civil whistleblower or *qui tam* actions, against individuals or entities for knowingly presenting, or causing to be presented, to the federal government, claims for payment that are false or fraudulent or making a false statement to avoid, decrease or conceal an obligation to pay money to the federal government. In addition, the government may assert that a claim including items and services resulting from a violation of the federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the False Claims Act. As a result of a modification made by the Fraud Enforcement and Recovery Act of 2009, a claim includes "any request or demand" for money or property presented to the federal government. In addition, manufacturers can be held liable under the 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;

- 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;
- HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act of 2009, or HITECH, and its implementing regulations, which also imposes obligations on certain covered entity healthcare providers, health plans and healthcare clearinghouses as well as their business associates that perform certain services involving the use or disclosure of individually identifiable health information, including mandatory contractual terms, with respect to safeguarding the privacy, security and transmission of individually identifiable health information; and
- analogous state laws and regulations, such as state anti-kickback and false claims laws that may apply to sales or marketing arrangements and claims involving healthcare items or services reimbursed by non-governmental third-party payors, including private insurers; and some state laws require pharmaceutical companies to comply with the pharmaceutical industry’s voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government in addition to requiring drug manufacturers to report information related to payments to physicians and other healthcare providers or marketing expenditures and pricing information, and state laws governing the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and often are not preempted by HIPAA, thus complicating compliance efforts.

These laws and regulations, among other things, may constrain our business, marketing and other promotional activities by limiting the kinds of financial arrangements we may have with hospitals, physicians or other potential purchasers of our product candidates, if approved. We have entered into consulting and advisory board agreements with physicians, some of whom are paid in the form of shares or options to acquire our common shares. We could be adversely affected if regulatory agencies determine our financial relationships with such physicians to be in violation of applicable laws 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 physicians are also governed by strict laws, regulations, industry self-regulation codes of conduct and physicians' codes of professional conduct in the individual EU member states. The provision of any inducements to physicians to prescribe, recommend, endorse, order, purchase, supply, use or administer a drug product is prohibited. A number of EU member states have established additional rules requiring pharmaceutical companies to publicly disclose their interactions with physicians and to obtain approval from employers, professional organizations or competent authorities before entering into agreements with physicians.

Ensuring that our future business arrangements with third parties comply with applicable healthcare laws and regulations will involve substantial costs. It is possible that governmental authorities will conclude that our business practices do not comply with current or future statutes, regulations or case law involving applicable fraud and abuse or other healthcare laws and regulations. If our operations, including anticipated activities to be conducted by our sales team, were to be found to be in violation of any of these laws or any other governmental regulations that may apply to us, we may be subject to the imposition of civil, criminal and administrative penalties, damages, disgorgement, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, individual imprisonment, contractual damages, reputational harm, diminished profits and future earnings, additional reporting requirements 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.

#### **Other Risks Related to Our Business**

***The COVID-19 pandemic, and measures taken in response to the pandemic, could adversely impact 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 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, could cause significant disruption in our business and operations as well as the business and operations of our manufacturers, CROs upon whom we rely 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. For example, in March 2020, the governors of Massachusetts and California, among other things, each enacted a stay-at-home advisory for workers in non-essential businesses. Because of the nature of our operations, we are currently considered to be an essential business so, to date, our operations have only been partially affected by this order. In response, we have temporarily closed access to our California office space and restricted access to our Massachusetts facility to only those employees that need to be in the office to execute their responsibilities and those employees who work in our research and development laboratory space, with most of our employees continuing to carry out their responsibilities working outside of our offices.

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, our manufacturers, CROs or other third parties with whom we conduct business or otherwise engage, may 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, has had an

impact on certain aspects of our commercialization strategy, including limiting interactions with third-party payors, physicians and patient advocacy groups to build disease awareness at industry conferences or otherwise, convening advisory boards and other research activities, 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 adversely impact our preclinical studies and clinical trials, which could 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 “The COVID-19 pandemic could adversely impact our preclinical studies and clinical trials.”

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 and social distancing, and the effectiveness of actions taken to contain and treat the disease.

***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 will face an even greater risk if we commercially sell any product candidates that we may develop. If we cannot successfully defend ourselves against claims that our product candidates caused injuries, we could incur substantial liabilities. Regardless of merit or eventual outcome, liability claims may result in:

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

Although we maintain product liability insurance coverage, it may not be adequate to cover all liabilities that we may incur and is subject to deductibles and coverage limitations. We anticipate that we will need to increase our insurance coverage when and if we successfully commercialize any product candidate. Insurance coverage is increasingly expensive. We may not be able to maintain insurance coverage at a reasonable cost or in an amount

adequate to satisfy any liability that may arise. If we are unable to obtain insurance at acceptable cost or otherwise protect against potential product liability claims, we will be exposed to significant liabilities, which may materially and adversely affect our business and financial position. These liabilities could prevent or interfere with our commercialization efforts.

***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 future operations. For example, in the United States, the Affordable Care Act substantially changes the way healthcare is financed by both governmental and private insurers, and significantly impacts the U.S. pharmaceutical industry. The Affordable Care Act, among other things, subjects biologic products to potential competition by lower-cost biosimilars, addresses a new methodology by which rebates owed by manufacturers under the Medicaid Drug Rebate Program are calculated for drugs and biologics that are inhaled, infused, instilled, implanted or injected, increases the minimum Medicaid rebates owed by manufacturers under the Medicaid Drug Rebate Program and extends the rebate program to individuals enrolled in Medicaid managed care organizations, establishes annual fees and taxes on manufacturers of certain branded prescription drugs and biologics, and a new Medicare Part D coverage gap discount program, in which manufacturers must agree to offer 50% point-of-sale discounts which, through subsequent legislative amendments, was increased to 70%, off negotiated prices of applicable brand drugs and biologics to eligible beneficiaries during their coverage gap period, as a condition for the manufacturer's outpatient products to be covered under Medicare Part D.

Since its enactment, there have been judicial and Congressional challenges to certain aspects of the Affordable Care Act. 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. It is unclear how these decisions, subsequent appeals, if any, or other efforts to challenge, repeal or replace the Affordable Care Act will impact the law or our business.

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 2029 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 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. 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, 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. 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 and the further development of our product candidates could be delayed.

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

Most healthcare providers, including research institutions from which we obtain patient health information, are subject to privacy and security regulations promulgated under HIPAA, as amended by the HITECH. We are not currently 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.

Our clinical trial programs outside the United States may implicate international data protection laws, including the EU Data Protection Directive and legislation of the EU member states implementing it. Our activities outside the United States impose additional compliance requirements and generate additional risks of enforcement for noncompliance. Failure by our CROs and other third-party contractors to comply with the strict rules on the transfer of personal data outside of the 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.

***The EU's data privacy regulation, the General Data Protection Regulation, has taken effect and violations of this could subject us to significant fines.***

In May 2018, a new privacy framework, the General Data Protection Regulation, or the GDPR, took effect in the EU and has become binding across all EEA member states. The GDPR imposes several stringent requirements for controllers and processors of personal data, particularly with respect to clinical trials. The GDPR provides that EU member states may make their own further laws and regulations limiting the processing of genetic, biometric or health data, which could limit our ability to use and share personal data or could cause our costs to increase and harm our business and financial condition. In addition, the GDPR increases the scrutiny that clinical trial sites located in the EEA should apply to transfers of personal data from such sites to countries that are considered to lack an adequate level of data protection, such as the United States. There are currently a number of legal challenges to the validity of EU mechanisms for adequate data transfers (such as the commonly-used EU-Commission-approved model clauses) or review of these mechanisms (such as the U.S. Privacy Shield), and our business could be impacted by changes in law as a result of a future review of these transfer mechanisms by EU regulators under the GDPR, as well as current challenges to these mechanisms in the EU courts. The GDPR imposes substantial fines for breaches of data protection requirements, which can be up to four percent of global revenue for the preceding financial year or €20 million, whichever is greater, and it also confers a private right of action on data subjects for breaches of data protection requirements. Compliance with EU data protection law is a rigorous and time-intensive process that may increase our cost of doing business, and despite those efforts, there is a risk that we may be subject to fines and penalties, litigation and reputational harm in connection with our European activities.

Additionally, following the United Kingdom's withdrawal from the European Union, we will have to comply with the GDPR and the United Kingdom GDPR, each regime having the ability to fine up to the greater of €20 million/ £17.5 million or 4% of global revenue. The relationship between the United Kingdom and the European Union in relation to certain aspects of data protection law remains unclear, for example around how data can lawfully be transferred between each jurisdiction, which exposes us to further compliance risk.

***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 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 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 Baker Bros. Advisors LP, or Baker Bros. Advisors, and Class B1 common shares, all of which shares are held by entities affiliated with Baker Bros. Advisors means that such persons are, and such entities may in the future be, able to influence or control 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, 2020, the holders of Class A common shares accounted for approximately 42% of our aggregate voting power and the holders of Class B common shares accounted for approximately 58% 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 52% of our aggregate voting power as of March 31, 2020 and have the ability to control 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, 2020, entities affiliated with Baker Bros. Advisors 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.

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.

This concentrated control limits certain shareholders' ability to influence corporate matters and 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 the Baker Bros. Advisors 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. These holders may have interests, with respect to their investment, that are different from our other shareholders. In addition, this concentration of control might adversely affect certain corporate actions that our other 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 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;
- the results and potential impact of competitive products or technologies;

- our ability to manufacture and successfully produce our product candidates;
- actual or anticipated changes in estimates as to financial results, 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;
- 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 affiliates who 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.

***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, 2020, upon such transfers or conversions up to approximately 34.1 million of additional Class A common shares would be 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, 2020, there were approximately 9.3 million Class A common shares subject to outstanding 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 funds affiliated with certain of our directors. As of March 31, 2020, on an as-converted basis, these shareholders collectively held approximately 31.9 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.

Further, as of (i) March 31, 2020, holders of approximately 35.0 million Class A common shares, including Class A common shares issuable upon the conversion 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, are entitled to certain rights with respect to the registration of these shares under the Securities Act pursuant to our amended and restated investor rights agreement, or the investors rights agreement and (ii) June 10, 2019, we registered 28,882,977 Class A common shares (inclusive of 3,000,000 Class A common shares acquired by certain of these holders in our initial public offering of our Class A common shares, or IPO) on an as converted basis from the registrable securities held by certain of these holders pursuant to the investors rights agreement, which are freely tradable without restriction under the Securities Act, to the extent permitted by Rule 144 under the Securities Act.

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. For example, we acquired the issued and outstanding equity securities of Primatope in exchange for upfront consideration of \$10.0 million paid at closing in March 2019 as well as milestone payments of \$5.0 million, which had been achieved and paid as of the closing date, and \$3.0 million which was paid in June 2019, all of which paid in a combination of cash and our Class A common shares (inclusive of escrow and holdback amounts) in accordance with the terms and conditions of our stock purchase option agreement with Primatope, or the “Primatope Agreement”.

***We are an “emerging growth company” and a “smaller reporting company” and the reduced disclosure requirements applicable to emerging growth companies and smaller reporting companies may make our Class A common shares less attractive to investors.***

We are an “emerging growth company,” as defined in the Jumpstart Our Business Startups, or JOBS, Act, and a “smaller reporting company” as defined under the rules promulgated under the Securities Act. As an emerging growth company and a smaller reporting company we may follow reduced disclosure requirements and do not have to make all of the disclosures that public companies that are not emerging growth companies or smaller reporting companies do. We will remain an emerging growth company until the earlier of:

- the last day of the fiscal year in which we have total annual gross revenues of \$1.07 billion or more;
- the last day of the fiscal year following the fifth anniversary of the date of the completion of our IPO;
- the date on which we have issued more than \$1 billion in nonconvertible debt during the previous three years; or
- the date on which we are deemed to be a large accelerated filer under the rules of the SEC, which means the market value of our voting and non-voting common shares that is held by non-affiliates exceeds \$700 million as of the last business day of our most recently completed second fiscal quarter.

For so long as we remain an emerging growth company, we are permitted and intend to rely on exemptions from certain disclosure requirements that are applicable to other public companies that are not emerging growth companies. These exemptions include:

- not being required to comply with the auditor attestation requirements of Section 404 of the Sarbanes-Oxley Act of 2002, or Section 404;
- an exemption from compliance with the requirement of the Public Company Accounting Oversight Board regarding the communication of critical audit matters in the auditor’s report on the financial statements;
- progressively adding to the number of years of audited financial statements required to be included in our periodic reports; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation, shareholder approval of any golden parachute payments not previously approved, and having to disclose the ratio of the compensation of our chief executive officer to the median compensation of our employees.

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

We are also a smaller reporting company, and we will remain a smaller reporting company until we no longer meet either of the criteria for being a smaller reporting company as follows:

- our voting and non-voting common shares held by non-affiliates is less than \$250 million measured on the last business day of our second fiscal quarter; or
- our annual revenues are less than \$100 million during the most recently completed fiscal year and our voting and non-voting common shares held by non-affiliates is less than \$700 million measured on the last business day of our second fiscal quarter.

Similar to emerging growth companies, smaller reporting companies are able to provide simplified executive compensation disclosure and have certain other reduced disclosure obligations, including, among other things, being required to provide only two years of audited financial statements and not being required to provide selected financial data, supplemental financial information or risk factors.

We may choose to take advantage of some, but not all, of the available exemptions for emerging growth companies and smaller reporting companies. We cannot predict whether investors will find our Class A common shares less attractive if we rely on these exemptions. If some investors find our Class A common shares less attractive as a result, there may be a less active trading market for our Class A common shares and the share price of our Class A common shares may be more volatile.

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

As a public company, 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, and will increase after we are no longer an emerging growth company and a smaller reporting company. For example, these rules and regulations may make it more difficult and more expensive for us to obtain director and officer liability insurance.

Pursuant to Section 404, we 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. However, while we remain an emerging growth company, we are not required to include an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. To maintain compliance with Section 404 within the prescribed period, 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<sup>2</sup>/<sub>3</sub>% 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 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 was to arise, it could adversely impact our tax position and our effective tax rate. Failure to manage the risks associated with such changes, or misinterpretation of the laws providing such changes, could result in costly audits, interest, penalties and reputational damage, which could adversely affect our business, results of 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.

In late 2017, the EU Economic and Financial Affairs Council, or ECOFIN, released a list of non-cooperative jurisdictions for tax purposes. The stated aim of this list, and accompanying report, was to promote the EU's view for good governance worldwide in order to maximize efforts to prevent tax fraud and tax evasion. While Bermuda was not on the original EU list of non-cooperative jurisdictions, it committed to address EU concerns relating to economic substance by December 31, 2018. In accordance with that commitment, Bermuda enacted the Economic Substance Act 2018, or the Substance Act, requiring certain entities in Bermuda engaged in "relevant activities" to maintain a substantial economic presence in Bermuda and to satisfy economic substance requirements commencing as of July 1, 2019. The list of "relevant activities" includes carrying on as a business in any one or more of: banking, insurance, fund management, financing, leasing, headquarters, shipping, distribution and service center, intellectual property and holding entities. Under the Substance Act, any entity that must satisfy economic substance requirements but fails to do so could face automatic disclosure to competent authorities in the EU of the information filed by the entity with the Bermuda Registrar of Companies in connection with the economic substance requirements and may also face financial penalties, restriction or regulation of its business activities or may be struck as a registered entity in Bermuda. Based on the results of our operations and guidance notes published by the Bermuda Minister of Finance on December 24, 2019, we believe that we are not currently in scope of the Substance Act as we do not believe we currently earn gross income from a "relevant activity," which is a condition to the application of the Substance Act. We will continue to monitor our status with respect to the Substance Act based on our results of operations, and may become subject to the Substance Act in future.

***We believe we will likely be classified as a passive foreign investment company for U.S. federal income tax purposes for the year ended December 31, 2020, which could result in adverse U.S. federal income tax consequences to U.S. investors in our common shares.***

Because we did not earn revenue from our business operations for the year ended December 31, 2019 and do not expect to do so for the year ended December 31, 2020, and because our sole source of income has been and currently is interest on bank accounts held by us, we believe we will be classified as a "passive foreign investment company," or PFIC, for the taxable year ended December 31, 2020. A non-U.S. company will be considered a PFIC for any taxable year if (i) at least 75% of its gross income is passive income (including interest income), or (ii) at least 50% of the value of its assets (based on an average of the quarterly values of the assets during a taxable year) is attributable to assets that produce or are held for the production of passive income. If we are classified as a PFIC in any year with respect to which a U.S. Holder 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 the U.S. Holder makes or has made a "qualified electing fund" election or a "mark-to-market" election and we cease to be a PFIC. 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 all or a portion of any gain realized on a disposition of our shares and on the receipt of distributions on our shares to the extent such gain or distribution is treated as an “excess distribution”, (ii) the application of a deferred interest charge 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, 2020. Even if we were not classified as a controlled foreign corporation, because our group includes one or more U.S. subsidiaries, certain of our non-U.S. subsidiaries could be treated as controlled foreign corporations. If a U.S. Holder is treated as owning (directly, indirectly or constructively) at least 10% of the value or voting power of common shares, such U.S. Holder may be treated as a “United States shareholder” with respect to us (if we are classified as a controlled foreign corporation) and each controlled foreign corporation in our group (if any). A United States shareholder of a controlled foreign corporation may be required to annually report and include in its U.S. taxable income its pro rata share of “Subpart F income,” “global intangible low-taxed income,” 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 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.

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

In 2017, the U.S. government enacted comprehensive tax legislation, known as the Tax Cuts and Jobs Act, or the TCJA, that included significant changes to the taxation of business entities. The TCJA remains unclear in many respects and has been, and may continue to be, the subject of amendments and technical corrections, as well as interpretations and implementing regulations by the Treasury and Internal Revenue Service, which have lessened or increased certain adverse impacts of the TCJA and may do so in the future. We continue to examine the impact the TCJA may have on our business. The effect of the TCJA on our business, whether adverse or favorable, is uncertain, and may not become evident for some period of time. Holders of our shares should consult with their legal and tax advisors regarding the TCJA, and any other such legislation, and the potential tax consequences of investing in our shares.

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

### ***Use of Proceeds from Registered Securities***

On May 29, 2018, we issued and sold 8,477,777 Class A common shares to the underwriters in our initial public offering, or IPO, and on June 22, 2018, we issued and sold an additional 1,006,425 Class A common shares pursuant to the exercise by the underwriters of their over-allotment option to purchase additional shares. Our Class A common shares were sold at a price to the public of \$18.00 per share. We received aggregate gross proceeds from the IPO inclusive of the underwriters’ over-allotment option of approximately \$170.7 million and aggregate net proceeds of approximately \$155.5 million after deducting underwriting discounts and commissions of approximately \$12.0 million and other offering expenses. The offer and sale of all of the shares in our IPO inclusive of the underwriters’ over-allotment option were registered under the Securities Act pursuant to a registration statement on Form S-1 (File No. 333-224488), which was declared effective by the Securities and Exchange Commission, or SEC, on May 23, 2018, and

a registration statement on Form S-1 to register additional shares (File No. 333-225159), which was automatically effective upon filing with the SEC on May 23, 2018. There has been no material change in our planned use of the net proceeds from the IPO as described in our final prospectus filed pursuant to Rule 424(b)(4) under the Securities Act with the SEC on May 24, 2018.

**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	<a href="#">Memorandum of Association of Kiniksa Pharmaceuticals, Ltd.</a>	S-1	333-224488	3.1	4/27/2018	
3.2	<a href="#">Amended and Restated Bye-Laws of Kiniksa Pharmaceuticals, Ltd.</a>	8-K	001-38492	3.1	5/29/2018	
4.1	<a href="#">Specimen Share Certificate evidencing the Class A common shares</a>	S-1/A	333-224488	4.1	5/14/2018	
4.2	<a href="#">Second Amended and Restated Investors' Rights Agreement, dated as of February 9, 2018</a>	S-1	333-224488	3.1	4/27/2018	
10.1	<a href="#">Form of Restricted Share Unit Grant Notice and Restricted Share Unit Agreement Thereunder for UK Participants under the 2018 Incentive Award plan; Sub-Plan for UK Employees</a>	S-8	333-237589	99.3	4/7/2020	
10.2	<a href="#">Forms of Performance Restricted Share Unit and Performance Cash Award Grant Notice and Performance Restricted Share Unit and Performance Cash Award Agreement thereunder and Restricted Share Unit Grant Notice and Restricted Share Unit Agreement thereunder for UK participants and German participants under the Rilonaconcept Long-Term Incentive Plan</a>	S-8	333-237589	99.8	4/7/2020	

Exhibit Number	Exhibit Description	Incorporated by Reference			Filed/ Furnished Herewith
		Form	File No.	Exhibit	
10.3	<a href="#">2018 Incentive Award Plan, and the form of share option grant notice and option agreement, form of restricted share grant notice and restricted share agreement, and form of restricted share unit grant notice and restricted share unit agreement thereunder</a>				*
31.1	<a href="#">Rule 13a-14(a) / 15d-14(a) Certification of Principal Executive Officer</a>				*
31.2	<a href="#">Rule 13a-14(a) / 15d-14(a) Certification of Principal Financial Officer</a>				*
32.1	<a href="#">Section 1350 Certification of Principal Executive Officer</a>				**
32.2	<a href="#">Section 1350 Certification of Principal Financial Officer</a>				**
101.INS	XBRL Instance Document				***
101.SCH	XBRL Taxonomy Extension Schema Document				***
101.CAL	XBRL Taxonomy Extension Calculation Linkbase Document				***
101.DEF	XBRL Extension Definition Linkbase Document				***
101.LAB	XBRL Taxonomy Label Linkbase Document				***
101.PRE	XBRL Taxonomy Extension Presentation Linkbase Document				***

\* 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 4, 2020

By: /s/ Sanj K. Patel

Sanj K. Patel

Chief Executive Officer and Chairman of the Board of Directors

**KINIKSA PHARMACEUTICALS, LTD.  
2018 INCENTIVE AWARD PLAN**

**ARTICLE I.  
PURPOSE**

The Plan's purpose is to enhance the Company's ability to attract, retain and motivate persons who make (or are expected to make) important contributions to the Company by providing these individuals with equity ownership opportunities. Capitalized terms used in the Plan are defined in Article XI.

**ARTICLE II.  
ELIGIBILITY**

Service Providers are eligible to be granted Awards under the Plan, subject to the limitations described herein.

**ARTICLE III.  
ADMINISTRATION AND DELEGATION**

3.1 Administration. The Plan is administered by the Administrator. The Administrator has authority to determine which Service Providers receive Awards, grant Awards and set Award terms and conditions, subject to the conditions and limitations in the Plan. The Administrator also has the authority to take all actions and make all determinations under the Plan, to interpret the Plan and Award Agreements and to adopt, amend and repeal Plan administrative rules, guidelines and practices as it deems advisable. The Administrator may correct defects and ambiguities, supply omissions and reconcile inconsistencies in the Plan or any Award as it deems necessary or appropriate to administer the Plan and any Awards. The Administrator's determinations under the Plan are in its sole discretion and will be final and binding on all persons having or claiming any interest in the Plan or any Award.

3.2 Appointment of Committees. To the extent Applicable Laws permit, the Board may delegate any or all of its powers under the Plan to one or more Committees or officers of the Company or any of its Subsidiaries. The Board may abolish any Committee or re-vest in itself any previously delegated authority at any time.

**ARTICLE IV.  
SHARES AVAILABLE FOR AWARDS**

4.1 Number of Shares. Subject to adjustment under Article VIII and the terms of this Article IV, Awards may be made under the Plan covering up to the Overall Share Limit. As of the Plan's effective date under Section 10.3, the Company will cease granting awards under the Prior Plans; however, Prior Plan Awards will remain subject to the terms of the applicable Prior Plan. Shares issued under the Plan may consist of authorized but unissued Shares, Shares purchased on the open market or Shares held in treasury.

4.2 Share Recycling. If all or any part of an Award or Prior Plan Award expires, lapses or is terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited, in any case, in a manner that results in the Company acquiring Shares covered by the Award or Prior Plan Award at a price not greater than the price (as adjusted to reflect any Equity Restructuring) paid by the Participant for such Shares or not issuing any Shares covered by the Award or Prior Plan Award, the unused Shares covered by the Award or Prior Plan Award will, as applicable, become or again be available

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for Award grants under the Plan. Further, Shares delivered (either by actual delivery or attestation) to the Company by a Participant to satisfy the applicable exercise or purchase price of an Award or Prior Plan Award and/or to satisfy any applicable tax withholding obligation, will, as applicable, become or again be available for Award grants under the Plan. The payment of Dividend Equivalents in cash in conjunction with any outstanding Awards shall not count against the Overall Share Limit.

4.3 Incentive Stock Option Limitations. Notwithstanding anything to the contrary herein, no more than 27,915,000 Shares may be issued pursuant to the exercise of Incentive Stock Options.

4.4 Substitute Awards. In connection with an entity's amalgamation, merger or consolidation with the Company or the Company's acquisition of an entity's property or shares, the Administrator may grant Awards in substitution for any options or other shares or share-based awards granted before such amalgamation, merger or consolidation by such entity or its affiliate. Substitute Awards may be granted on such terms as the Administrator deems appropriate, notwithstanding limitations on Awards in the Plan. Substitute Awards will not count against the Overall Share Limit (nor shall Shares subject to a Substitute Award be added to the Shares available for Awards under the Plan as provided above), except that Shares acquired by exercise of substitute Incentive Stock Options will count against the maximum number of Shares that may be issued pursuant to the exercise of Incentive Stock Options under the Plan. Additionally, in the event that a company acquired by the Company or any Subsidiary or with which the Company or any Subsidiary combines has shares available under a pre-existing plan approved by shareholders and not adopted in contemplation of such acquisition or combination, the shares available for grant pursuant to the terms of such pre-existing plan (as adjusted, to the extent appropriate, using the exchange ratio or other adjustment or valuation ratio or formula used in such acquisition or combination to determine the consideration payable to the equity holders of the entities party to such acquisition or combination) may be used for Awards under the Plan and shall not reduce the Shares authorized for grant under the Plan (and Shares subject to such Awards shall not be added to the Shares available for Awards under the Plan as provided above); provided that Awards using such available shares shall not be made after the date awards or grants could have been made under the terms of the pre-existing plan, absent the acquisition or combination, and shall only be made to individuals who were not Employees or Directors prior to such acquisition or combination.

## **ARTICLE V. SHARE OPTIONS AND SHARE APPRECIATION RIGHTS**

5.1 General. The Administrator may grant Options or Share Appreciation Rights to Service Providers subject to the limitations in the Plan, including any limitations in the Plan that apply to Incentive Stock Options. The Administrator will determine the number of Shares covered by each Option and Share Appreciation Right, the exercise price of each Option and Share Appreciation Right, the vesting conditions of each Option and Share Appreciation Right, and the conditions and limitations applicable to the exercise of each Option and Share Appreciation Right. A Share Appreciation Right will entitle the Participant (or other person entitled to exercise the Share Appreciation Right) to receive from the Company upon exercise of the exercisable portion of the Share Appreciation Right an amount determined by multiplying the excess, if any, of the Fair Market Value of one Share on the date of exercise over the exercise price per Share of the Share Appreciation Right by the number of Shares with respect to which the Share Appreciation Right is exercised, subject to any limitations of the Plan or that the Administrator may impose and payable in cash, Shares valued at Fair Market Value or a combination of the two as the Administrator may determine or provide in the Award Agreement.

5.2 Exercise Price. The Administrator will establish each Option's and Share Appreciation Right's exercise price and specify the exercise price in the Award Agreement. The exercise price will not

be less than 100% of the Fair Market Value or par value per Share, whichever is greater, on the grant date of the Option or Share Appreciation Right.

5.3 Duration. Each Option or Share Appreciation Right will be exercisable at such times and as specified in the Award Agreement, provided that the term of an Option or Share Appreciation Right will not exceed ten years. Notwithstanding the foregoing and unless determined otherwise by the Company, in the event that on the last business day of the term of an Option or Share Appreciation Right (other than an Incentive Stock Option) (i) the exercise of the Option or Share Appreciation Right is prohibited by Applicable Law, as determined by the Company, or (ii) Shares may not be purchased or sold by the applicable Participant due to any Company insider trading policy (including blackout periods) or a “lock-up” agreement undertaken in connection with an issuance of securities by the Company, the term of the Option or Share Appreciation Right shall be extended until the date that is thirty (30) days after the end of the legal prohibition, black-out period or lock-up agreement, as determined by the Company; provided, however, in no event shall the extension last beyond the ten year term of the applicable Option or Share Appreciation Right. Notwithstanding the foregoing, if the Participant, prior to the end of the term of an Option or Share Appreciation Right, violates the non-competition, non-solicitation, confidentiality or other similar restrictive covenant provisions of any employment contract, confidentiality and nondisclosure agreement or other agreement between the Participant and the Company or any of its Subsidiaries, the right of the Participant and the Participant’s transferees to exercise any Option or Share Appreciation Right issued to the Participant shall terminate immediately upon such violation, unless the Company otherwise determines. In addition, if, prior to the end of the term of an Option or Share Appreciation Right, the Participant is given notice by the Company or any of its Subsidiaries of the Participant’s Termination of Service by the Company or any of its Subsidiaries for Cause, and the effective date of such Termination of Service is subsequent to the date of the delivery of such notice, the right of the Participant and the Participant’s transferees to exercise any Option or Share Appreciation Right issued to the Participant shall be suspended from the time of the delivery of such notice until the earlier of (i) such time as it is determined or otherwise agreed that the Participant’s service as a Service Provider will not be terminated for Cause as provided in such notice or (ii) the effective date of the Participant’s Termination of Service by the Company or any of its Subsidiaries for Cause (in which case the right of the Participant and the Participant’s transferees to exercise any Option or Share Appreciation Right issued to the Participant will terminate immediately upon the effective date of such termination of Service).

5.4 Exercise. Options and Share Appreciation Rights may be exercised by delivering to the Company a written notice of exercise, in a form the Administrator approves (which may be electronic), signed by the person authorized to exercise the Option or Share Appreciation Right, together with, as applicable, payment in full (i) as specified in Section 5.5 for the number of Shares for which the Award is exercised and (ii) as specified in Section 9.5 for any applicable taxes. Unless the Administrator otherwise determines, an Option or Share Appreciation Right may not be exercised for a fraction of a Share.

5.5 Payment Upon Exercise. Subject to Section 10.8, any Company insider trading policy (including blackout periods) and Applicable Laws, the exercise price of an Option must be paid by:

(a) cash, wire transfer of immediately available funds or by check payable to the order of the Company, provided that the Company may limit the use of one of the foregoing payment forms if one or more of the payment forms below is permitted;

(b) if there is a public market for Shares at the time of exercise, unless the Company otherwise determines, (A) delivery (including telephonically to the extent permitted by the Company) of an irrevocable and unconditional undertaking by a broker acceptable to the Company to deliver promptly to the Company sufficient funds to pay the exercise price, or (B) the Participant’s delivery to the Company of a copy of irrevocable and unconditional instructions to a broker acceptable to the Company to deliver

promptly to the Company cash or a check sufficient to pay the exercise price; provided that such amount is paid to the Company at such time as may be required by the Administrator;

(c) to the extent permitted by the Administrator, delivery (either by actual delivery or attestation) of Shares owned by the Participant valued at their Fair Market Value;

(d) to the extent permitted by the Administrator, surrendering Shares then issuable upon the Option's exercise valued at their Fair Market Value on the exercise date;

(e) to the extent permitted by the Administrator, delivery of a promissory note or any other property that the Administrator determines is good and valuable consideration; or

(f) to the extent permitted by the Company, any combination of the above payment forms approved by the Administrator.

## **ARTICLE VI. RESTRICTED SHARES; RESTRICTED SHARE UNITS**

6.1 General. The Administrator may grant Restricted Shares, or the right to purchase Restricted Shares, to any Service Provider, subject to the Company's right to repurchase all or part of such shares at their issue price or other stated or formula price from the Participant (or to require forfeiture of such shares) if conditions the Administrator specifies in the Award Agreement are not satisfied before the end of the applicable restriction period or periods that the Administrator establishes for such Award and subject to Applicable Laws. In addition, the Administrator may grant to Service Providers Restricted Share Units, which may be subject to vesting and forfeiture conditions during the applicable restriction period or periods, as set forth in an Award Agreement. The Administrator will determine and set forth in the Award Agreement the terms and conditions for each Restricted Share and Restricted Share Unit Award, subject to the conditions and limitations contained in the Plan.

### 6.2 Restricted Shares.

(a) Dividends. Participants holding shares of Restricted Shares will be entitled to all ordinary cash dividends paid with respect to such Shares, unless the Administrator provides otherwise in the Award Agreement. In addition, unless the Administrator provides otherwise, if any dividends or distributions are paid in Shares, or consist of a dividend or distribution to holders of Common Shares of property other than an ordinary cash dividend, the Shares or other property will be subject to the same restrictions on transferability and forfeitability as the shares of Restricted Shares with respect to which they were paid.

(b) Share Certificates. The Company may require that the Participant deposit in escrow with the Company (or its designee) any share certificates issued in respect of shares of Restricted Shares, together with a duly executed, but undated, instrument of transfer.

### 6.3 Restricted Share Units.

(a) Settlement. The Administrator may provide that settlement of Restricted Share Units will occur upon or as soon as reasonably practicable after the Restricted Share Units vest or will instead be deferred, on a mandatory basis or at the Participant's election, in a manner intended to comply with Section 409A.

(b) Shareholder Rights. A Participant will have no rights of a shareholder with respect to Shares subject to any Restricted Share Unit unless and until the Shares are delivered in settlement of the Restricted Share Unit.

(c) Dividend Equivalents. If the Administrator provides, a grant of Restricted Share Units may provide a Participant with the right to receive Dividend Equivalents. Dividend Equivalents may be paid currently or credited to an account for the Participant, settled in cash or Shares and subject to the same restrictions on transferability and forfeitability as the Restricted Share Units with respect to which the Dividend Equivalents are granted and subject to other terms and conditions as set forth in the Award Agreement.

**ARTICLE VII.  
OTHER SHARE OR CASH BASED AWARDS**

Other Share or Cash Based Awards may be granted to Participants, including Awards entitling Participants to receive Shares to be delivered in the future and including annual or other periodic or long-term cash bonus awards (whether based on specified Performance Criteria or otherwise), in each case subject to any conditions and limitations in the Plan. Such Other Share or Cash Based Awards will also be available as a payment form in the settlement of other Awards, as standalone payments and as payment in lieu of compensation to which a Participant is otherwise entitled. Other Share or Cash Based Awards may be paid in Shares, cash or other property, as the Administrator determines. Subject to the provisions of the Plan, the Administrator will determine the terms and conditions of each Other Share or Cash Based Award, including any purchase price, performance goal (which may be based on the Performance Criteria), transfer restrictions, and vesting conditions, which will be set forth in the applicable Award Agreement.

**ARTICLE VIII.  
ADJUSTMENTS FOR CHANGES IN COMMON SHARES  
AND CERTAIN OTHER EVENTS**

8.1 Equity Restructuring. In connection with any Equity Restructuring, notwithstanding anything to the contrary in this Article VIII, the Administrator will equitably adjust each outstanding Award as it deems appropriate to reflect the Equity Restructuring, which may include adjusting the number and type of securities subject to each outstanding Award and/or the Award's exercise price or grant price (if applicable), granting new Awards to Participants, and making a cash payment to Participants. The adjustments provided under this Section 8.1 will be nondiscretionary and final and binding on the affected Participant and the Company; provided that the Administrator will determine whether an adjustment is equitable.

8.2 Corporate Transactions. In the event of any dividend or other distribution (whether in the form of cash, Common Shares, other securities, or other property), reorganization, merger, consolidation, combination, amalgamation, repurchase, recapitalization, liquidation, dissolution, or sale, transfer, exchange or other disposition of all or substantially all of the assets of the Company, or sale or exchange of Common Shares or other securities of the Company, Change in Control, issuance of warrants or other rights to purchase Common Shares or other securities of the Company, other similar corporate transaction or event, other unusual or nonrecurring transaction or event affecting the Company or its financial statements or any change in any Applicable Laws or accounting principles, the Administrator, on such terms and conditions as it deems appropriate, either by the terms of the Award or by action taken prior to the occurrence of such transaction or event (except that action to give effect to a change in Applicable Law or accounting principles may be made within a reasonable period of time after such change) and either automatically or upon the Participant's request, is hereby authorized to take any one or more of the following actions whenever the Administrator determines that such action is appropriate in order to (x)

prevent dilution or enlargement of the benefits or potential benefits intended by the Company to be made available under the Plan or with respect to any Award granted or issued under the Plan, (y) to facilitate such transaction or event or (z) give effect to such changes in Applicable Laws or accounting principles:

(a) To provide for the cancellation of any such Award in exchange for either an amount of cash or other property with a value equal to the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participant's rights under the vested portion of such Award, as applicable; provided that, if the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participant's rights, in any case, is equal to or less than zero, then the Award may be terminated without payment;

(b) To provide that such Award shall vest and, to the extent applicable, be exercisable as to all shares covered thereby, notwithstanding anything to the contrary in the Plan or the provisions of such Award;

(c) To provide that such Award be assumed by the successor or survivor corporation, or a parent or subsidiary thereof, or shall be substituted for by awards covering the shares of the successor or survivor corporation, or a parent or subsidiary thereof, with appropriate adjustments as to the number and kind of shares and/or applicable exercise or purchase price, in all cases, as determined by the Administrator;

(d) To make adjustments in the number and type of shares (or other securities or property) subject to outstanding Awards and/or with respect to which Awards may be granted under the Plan (including, but not limited to, adjustments of the limitations in Article IV hereof on the maximum number and kind of shares which may be issued) and/or in the terms and conditions of (including the grant or exercise price), and the criteria included in, outstanding Awards;

(e) To replace such Award with other rights or property selected by the Administrator; and/or

(f) To provide that the Award will terminate and cannot vest, be exercised or become payable after the applicable event.

8.3 Administrative Stand Still. In the event of any pending share dividend, bonus issue, share split, combination or exchange of shares, merger, amalgamation, consolidation or other distribution (other than normal cash dividends) of Company assets to shareholders, or any other extraordinary transaction or change affecting the Shares or the share price of Common Shares, including any Equity Restructuring or any securities offering or other similar transaction, for administrative convenience, the Administrator may refuse to permit the exercise of any Award for up to sixty days before or after such transaction.

8.4 General. Except as expressly provided in the Plan or the Administrator's action under the Plan, no Participant will have any rights due to any subdivision or consolidation of Shares of any class, dividend payment, increase or decrease in the number of Shares of any class or dissolution, liquidation, merger, amalgamation or consolidation of the Company or other corporation. Except as expressly provided with respect to an Equity Restructuring under Section 8.1 above or the Administrator's action under the Plan, no issuance by the Company of Shares of any class, or securities convertible into Shares of any class, will affect, and no adjustment will be made regarding, the number of Shares subject to an Award or the Award's grant or exercise price. The existence of the Plan, any Award Agreements and the Awards granted hereunder will not affect or restrict in any way the Company's right or power to make or authorize (i) any adjustment, recapitalization, reorganization or other change in the Company's capital structure or its business, (ii) any merger, amalgamation, consolidation dissolution or liquidation of the Company or sale of

Company assets or (iii) any sale or issuance of securities, including securities with rights superior to those of the Shares or securities convertible into or exchangeable for Shares. The Administrator may treat Participants and Awards (or portions thereof) differently under this Article VIII.

**ARTICLE IX.**  
**GENERAL PROVISIONS APPLICABLE TO AWARDS**

9.1 Transferability. Except as the Administrator may determine or provide in an Award Agreement or otherwise for Awards other than Incentive Stock Options, Awards may not be sold, assigned, transferred, pledged or otherwise encumbered, either voluntarily or by operation of law, except by will or the laws of descent and distribution, or, subject to the Administrator's consent, pursuant to a domestic relations order, and, during the life of the Participant, will be exercisable only by the Participant. References to a Participant, to the extent relevant in the context, will include references to a Participant's authorized transferee that the Administrator specifically approves.

9.2 Documentation. Each Award will be evidenced in an Award Agreement, which may be written or electronic, as the Administrator determines. Each Award may contain terms and conditions in addition to those set forth in the Plan.

9.3 Discretion. Except as the Plan otherwise provides, each Award may be made alone or in addition or in relation to any other Award. The terms of each Award to a Participant need not be identical, and the Administrator need not treat Participants or Awards (or portions thereof) uniformly.

9.4 Termination of Status. The Administrator will determine how the disability, death, retirement, authorized leave of absence or any other change or purported change in a Participant's Service Provider status affects an Award and the extent to which, and the period during which, the Participant, the Participant's legal representative, conservator, guardian or Designated Beneficiary may exercise rights under the Award, if applicable.

9.5 Withholding. Each Participant must pay the Company, or make provision satisfactory to the Administrator for payment of, any taxes required by law to be withheld in connection with such Participant's Awards by the date of the event creating the tax liability. The Company may deduct a cash amount sufficient to satisfy such tax obligations based on the applicable statutory withholding rates (or such other rate as may be determined by the Company after considering any accounting consequences or costs) from any payment otherwise due to a Participant. Subject to Section 10.8 and any Company insider trading policy (including blackout periods), Participants may satisfy such tax obligations (i) in cash, by wire transfer of immediately available funds, by check made payable to the order of the Company, provided that the Company may limit the use of the foregoing payment forms if one or more of the payment forms below is permitted, (ii) to the extent permitted by the Administrator, in whole or in part by delivery of Shares valued at their Fair Market Value or elect to have the Company repurchase Shares otherwise issuable under an Award limited to the number of Common Shares which have a Fair Market Value on the date of repurchase necessary to pay the aggregate amount of tax liability, (iii) if there is a public market for Shares at the time the tax obligations are satisfied, unless the Company otherwise determines, (A) delivery (including telephonically to the extent permitted by the Company) of an irrevocable and unconditional undertaking by a broker acceptable to the Company to deliver promptly to the Company sufficient funds to satisfy the tax obligations, or (B) delivery by the Participant to the Company of a copy of irrevocable and unconditional instructions to a broker acceptable to the Company to deliver promptly to the Company cash or a check sufficient to satisfy the tax withholding; provided that such amount is paid to the Company at such time as may be required by the Administrator, or (iv) to the extent permitted by the Company, any combination of the foregoing payment forms approved by the Administrator. If any tax withholding obligation will be satisfied under clause (ii) of the immediately preceding sentence by the Company's retention of Shares

from the Award creating the tax obligation and there is a public market for Shares at the time the tax obligation is satisfied, the Company may elect to instruct any brokerage firm determined acceptable to the Company for such purpose to sell on the applicable Participant's behalf some or all of the Shares retained and to remit the proceeds of the sale to the Company or its designee, and each Participant's acceptance of an Award under the Plan will constitute the Participant's authorization to the Company and instruction and authorization to such brokerage firm to complete the transactions described in this sentence.

9.6 Amendment of Award; Repricing. The Administrator may amend, modify or terminate any outstanding Award, including by substituting another Award of the same or a different type, changing the exercise or settlement date, and converting an Incentive Stock Option to a Non-Qualified Share Option. The Participant's consent to such action will be required unless (i) the action, taking into account any related action, does not materially and adversely affect the Participant's rights under the Award, or (ii) the change is permitted under Article VIII or pursuant to Section 10.6. Notwithstanding the foregoing or anything in the Plan to the contrary, the Administrator may not except pursuant to Article VIII, without the approval of the shareholders of the Company, reduce the exercise price per share of outstanding Options or Share Appreciation Rights or cancel outstanding Options or Share Appreciation Rights in exchange for cash, other Awards or Options or Share Appreciation Rights with an exercise price per share that is less than the exercise price per share of the original Options or Share Appreciation Rights.

9.7 Conditions on Delivery of Shares. The Company will not be obligated to deliver any Shares under the Plan or remove restrictions from Shares previously delivered under the Plan until (i) all Award conditions have been met or removed to the Company's satisfaction, (ii) as determined by the Company, all other legal matters regarding the issuance and delivery of such Shares have been satisfied, including any applicable securities laws and stock exchange or stock market rules and regulations, and (iii) the Participant has executed and delivered to the Company such representations or agreements as the Administrator deems necessary or appropriate to satisfy any Applicable Laws. The Company's inability to obtain authority from any regulatory body having jurisdiction, which the Administrator determines is necessary to the lawful issuance and sale of any securities, will relieve the Company of any liability for failing to issue or sell such Shares as to which such requisite authority has not been obtained.

9.8 Acceleration. The Administrator may at any time provide that any Award will become immediately vested and fully or partially exercisable, free of some or all restrictions or conditions, or otherwise fully or partially realizable.

9.9 Additional Terms of Incentive Stock Options. The Administrator may grant Incentive Stock Options only to employees of the Company, any of its present or future parent or subsidiary corporations, as defined in Sections 424(e) or (f) of the Code, respectively, and any other entities the employees of which are eligible to receive Incentive Stock Options under the Code. If an Incentive Stock Option is granted to a Greater Than 10% Shareholder, the exercise price will not be less than 110% of the Fair Market Value on the Option's grant date, and the term of the Option will not exceed five years. All Incentive Stock Options will be subject to and construed consistently with Section 422 of the Code. By accepting an Incentive Stock Option, the Participant agrees to give prompt notice to the Company of dispositions or other transfers (other than in connection with a Change in Control) of Shares acquired under the Option made within (i) two years from the grant date of the Option or (ii) one year after the transfer of such Shares to the Participant, specifying the date of the disposition or other transfer and the amount the Participant realized, in cash, other property, assumption of indebtedness or other consideration, in such disposition or other transfer. Neither the Company nor the Administrator will be liable to a Participant, or any other party, if an Incentive Stock Option fails or ceases to qualify as an "incentive stock option" under Section 422 of the Code. Any Incentive Stock Option or portion thereof that fails to qualify as an "incentive stock option" under Section 422 of the Code for any reason, including becoming exercisable with respect

to Shares having a fair market value exceeding the \$100,000 limitation under Treasury Regulation Section 1.422-4, will be a Non-Qualified Stock Option.

**ARTICLE X.  
MISCELLANEOUS**

10.1 No Right to Employment or Other Status. No person will have any claim or right to be granted an Award, and the grant of an Award will not be construed as giving a Participant the right to continued employment or any other relationship with the Company. The Company expressly reserves the right at any time to dismiss or otherwise terminate its relationship with a Participant free from any liability or claim under the Plan or any Award, except as expressly provided in an Award Agreement.

10.2 No Rights as Shareholder; Certificates. Subject to the Award Agreement, no Participant or Designated Beneficiary will have any rights as a shareholder with respect to any Shares to be distributed under an Award until becoming the record holder of such Shares. Notwithstanding any other provision of the Plan, unless the Administrator otherwise determines or Applicable Laws require, the Company will not be required to deliver to any Participant certificates evidencing Shares issued in connection with any Award and instead such Shares may be recorded in the books of the Company (or, as applicable, its transfer agent or share plan administrator). The Company may place legends on share certificates issued under the Plan that the Administrator deems necessary or appropriate to comply with Applicable Laws.

10.3 Effective Date and Term of Plan. Unless earlier terminated by the Board, the Plan will become effective on the day prior to the Public Trading Date and will remain in effect until the tenth anniversary of the earlier of (i) the date the Board adopted the Plan or (ii) the date the Company's shareholders approved the Plan, but Awards previously granted may extend beyond that date in accordance with the Plan. If the Plan is not approved by the Company's shareholders, the Plan will not become effective, no Awards will be granted under the Plan and the Prior Plans will continue in full force and effect in accordance with their terms.

10.4 Amendment of Plan. The Administrator may amend, suspend or terminate the Plan at any time; provided that no amendment, other than an increase to the Overall Share Limit, may materially and adversely affect any Award outstanding at the time of such amendment without the affected Participant's consent. No Awards may be granted under the Plan during any suspension period or after Plan termination. Awards outstanding at the time of any Plan suspension or termination will continue to be governed by the Plan and the Award Agreement, as in effect before such suspension or termination. The Board will obtain shareholder approval of any Plan amendment to the extent necessary to comply with Applicable Laws.

10.5 Provisions for Non-U.S. Employees. The Administrator may modify Awards granted to Participants who are citizens or residents of a country other than the United States or employed outside the United States or establish subplans or procedures under the Plan to address differences in laws, rules, regulations or customs of such foreign jurisdictions with respect to tax, securities, currency, employee benefit or other matters.

10.6 Section 409A.

(a) General. The Company intends that all Awards be structured to comply with, or be exempt from, Section 409A, such that no adverse tax consequences, interest, or penalties under Section 409A apply. Notwithstanding anything in the Plan or any Award Agreement to the contrary, the Administrator may, without a Participant's consent, amend this Plan or Awards, adopt policies and procedures, or take any other actions (including amendments, policies, procedures and retroactive actions) as are necessary or appropriate to preserve the intended tax treatment of Awards, including any such actions

intended to (A) exempt this Plan or any Award from Section 409A, or (B) comply with Section 409A, including regulations, guidance, compliance programs and other interpretative authority that may be issued after an Award's grant date. The Company makes no representations or warranties as to an Award's tax treatment under Section 409A or otherwise. The Company will have no obligation under this Section 10.6 or otherwise to avoid the taxes, penalties or interest under Section 409A with respect to any Award and will have no liability to any Participant or any other person if any Award, compensation or other benefits under the Plan are determined to constitute noncompliant "nonqualified deferred compensation" subject to taxes, penalties or interest under Section 409A.

(b) Separation from Service. If an Award constitutes "nonqualified deferred compensation" under Section 409A, any payment or settlement of such Award upon a termination of a Participant's Service Provider relationship will, to the extent necessary to avoid taxes under Section 409A, be made only upon the Participant's "separation from service" (within the meaning of Section 409A), whether such "separation from service" occurs upon or after the termination of the Participant's Service Provider relationship. For purposes of this Plan or any Award Agreement relating to any such payments or benefits, references to a "termination," "termination of employment" or like terms means a "separation from service."

(c) Payments to Specified Employees. Notwithstanding any contrary provision in the Plan or any Award Agreement, any payment(s) of "nonqualified deferred compensation" required to be made under an Award to a "specified employee" (as defined under Section 409A and as the Administrator determines) due to his or her "separation from service" will, to the extent necessary to avoid taxes under Section 409A(a)(2)(B)(i) of the Code, be delayed for the six-month period immediately following such "separation from service" (or, if earlier, until the specified employee's death) and will instead be paid (as set forth in the Award Agreement) on the day immediately following such six-month period or as soon as administratively practicable thereafter (without interest). Any payments of "nonqualified deferred compensation" under such Award payable more than six months following the Participant's "separation from service" will be paid at the time or times the payments are otherwise scheduled to be made.

10.7 Limitations on Liability. Notwithstanding any other provisions of the Plan, no individual acting as a director, officer, other employee or agent of the Company or any Subsidiary will be liable to any Participant, former Participant, spouse, beneficiary, or any other person for any claim, loss, liability, or expense incurred in connection with the Plan or any Award, and such individual will not be personally liable with respect to the Plan because of any contract or other instrument executed in his or her capacity as an Administrator, director, officer, other employee or agent of the Company or any Subsidiary. The Company will indemnify and hold harmless each director, officer, other employee and agent of the Company or any Subsidiary that has been or will be granted or delegated any duty or power relating to the Plan's administration or interpretation, against any cost or expense (including attorneys' fees) or liability (including any sum paid in settlement of a claim with the Administrator's approval) arising from any act or omission concerning this Plan unless arising from such person's own fraud or bad faith.

10.8 Lock-Up Period. The Company may, at the request of any underwriter representative or otherwise, in connection with registering the offering of any Company securities under the Securities Act, prohibit Participants from, directly or indirectly, selling or otherwise transferring any Shares or other Company securities during a period of up to one hundred eighty days following the effective date of a Company registration statement filed under the Securities Act, or such longer period as determined by the underwriter.

10.9 Data Privacy. As a condition for receiving any Award, each Participant explicitly and unambiguously consents to the collection, use and transfer, in electronic or other form, of personal data as described in this section by and among the Company and its Subsidiaries and affiliates exclusively for

implementing, administering and managing the Participant's participation in the Plan. The Company and its Subsidiaries and affiliates may hold certain personal information about a Participant, including the Participant's name, address and telephone number; birthdate; social security, insurance number or other identification number; salary; nationality; job title(s); any Shares held in the Company or its Subsidiaries and affiliates; and Award details, to implement, manage and administer the Plan and Awards (the "**Data**"). The Company and its Subsidiaries and affiliates may transfer the Data amongst themselves as necessary to implement, administer and manage a Participant's participation in the Plan, and the Company and its Subsidiaries and affiliates may transfer the Data to third parties assisting the Company with Plan implementation, administration and management. These recipients may be located in the Participant's country, or elsewhere, and the Participant's country may have different data privacy laws and protections than the recipients' country. By accepting an Award, each Participant authorizes such recipients to receive, possess, use, retain and transfer the Data, in electronic or other form, to implement, administer and manage the Participant's participation in the Plan, including any required Data transfer to a broker or other third party with whom the Company or the Participant may elect to deposit any Shares. The Data related to a Participant will be held only as long as necessary to implement, administer, and manage the Participant's participation in the Plan. A Participant may, at any time, view the Data that the Company holds regarding such Participant, request additional information about the storage and processing of the Data regarding such Participant, recommend any necessary corrections to the Data regarding the Participant or refuse or withdraw the consents in this Section 10.9 in writing, without cost, by contacting the local human resources representative. The Company may cancel Participant's ability to participate in the Plan and, in the Administrator's discretion, the Participant may forfeit any outstanding Awards if the Participant refuses or withdraws the consents in this Section 10.9. For more information on the consequences of refusing or withdrawing consent, Participants may contact their local human resources representative.

10.10 Severability. If any portion of the Plan or any action taken under it is held illegal or invalid for any reason, the illegality or invalidity will not affect the remaining parts of the Plan, and the Plan will be construed and enforced as if the illegal or invalid provisions had been excluded, and the illegal or invalid action will be null and void.

10.11 Governing Documents. If any contradiction occurs between the Plan and any Award Agreement or other written agreement between a Participant and the Company (or any Subsidiary) that the Administrator has approved, the Plan will govern, unless it is expressly specified in such Award Agreement or other written document that a specific provision of the Plan will not apply.

10.12 Governing Law. The Plan and all Awards will be governed by and interpreted in accordance with the laws of Delaware, disregarding any state's choice-of-law principles requiring the application of a jurisdiction's laws other than Delaware.

10.13 Claw-back Provisions. All Awards (including any proceeds, gains or other economic benefit the Participant actually or constructively receives upon receipt or exercise of any Award or the receipt or resale of any Shares underlying the Award) will be subject to any Company claw-back policy, including any claw-back policy adopted to comply with Applicable Laws (including the Dodd-Frank Wall Street Reform and Consumer Protection Act and any rules or regulations promulgated thereunder) as set forth in such claw-back policy or the Award Agreement.

10.14 Titles and Headings. The titles and headings in the Plan are for convenience of reference only and, if any conflict, the Plan's text, rather than such titles or headings, will control.

10.15 Conformity to Securities Laws. Participant acknowledges that the Plan is intended to conform to the extent necessary with Applicable Laws. Notwithstanding anything herein to the contrary, the Plan and all Awards will be administered only in conformance with Applicable Laws. To the extent

Applicable Laws permit, the Plan and all Award Agreements will be deemed amended as necessary to conform to Applicable Laws.

10.16 Relationship to Other Benefits. No payment under the Plan will be taken into account in determining any benefits under any pension, retirement, savings, profit sharing, group insurance, welfare or other benefit plan of the Company or any Subsidiary except as expressly provided in writing in such other plan or an agreement thereunder.

10.17 Broker-Assisted Sales. In the event of a broker-assisted sale of Shares in connection with the payment of amounts owed by a Participant under or with respect to the Plan or Awards, including amounts to be paid under the final sentence of Section 9.5: (a) any Shares to be sold through the broker-assisted sale will be sold on the day the payment first becomes due, or as soon thereafter as practicable; (b) such Shares may be sold as part of a block trade with other Participants in the Plan in which all participants receive an average price; (c) the applicable Participant will be responsible for all broker's fees and other costs of sale, and by accepting an Award, each Participant agrees to indemnify and hold the Company harmless from any losses, costs, damages, or expenses relating to any such sale; (d) to the extent the Company or its designee receives proceeds of such sale that exceed the amount owed, the Company will pay such excess in cash to the applicable Participant as soon as reasonably practicable; (e) the Company and its designees are under no obligation to arrange for such sale at any particular price; and (f) in the event the proceeds of such sale are insufficient to satisfy the Participant's applicable obligation, the Participant may be required to pay immediately upon demand to the Company or its designee an amount in cash sufficient to satisfy any remaining portion of the Participant's obligation.

## ARTICLE XI. DEFINITIONS

As used in the Plan, the following words and phrases will have the following meanings:

11.1 **"Administrator"** means the Board or a Committee to the extent that the Board's powers or authority under the Plan have been delegated to such Committee.

11.2 **"Applicable Laws"** means the requirements relating to the administration of equity incentive plans under U.S. federal and state securities, tax and other applicable laws, rules and regulations, the applicable rules of any stock exchange or quotation system on which the Common Shares are listed or quoted and the applicable laws and rules of any foreign country or other jurisdiction where Awards are granted or issued under the Plan, including without limitation, the laws of Bermuda.

11.3 **"Award"** means, individually or collectively, a grant under the Plan of Options, Share Appreciation Rights, Restricted Shares, Restricted Share Units or Other Share or Cash Based Awards.

11.4 **"Award Agreement"** means a written agreement evidencing an Award, which may be electronic, that contains such terms and conditions as the Administrator determines, consistent with and subject to the terms and conditions of the Plan.

11.5 **"Board"** means the Board of Directors of the Company.

11.6 **"Cause"** means, with respect to a Participant, (A) dishonesty with respect to the Company or any Subsidiary; (B) insubordination, substantial malfeasance or nonfeasance of duty; (C) unauthorized disclosure of confidential information; (D) breach by a Participant of any provision of any employment, consulting, advisory, nondisclosure, non-competition or similar agreement between the Participant and the Company or any Subsidiary; and (E) conduct substantially prejudicial to the business of the Company or

any Subsidiary; provided, however, that any provision in an agreement between a Participant and the Company or an Subsidiary, which contains a conflicting definition of Cause for termination and which is in effect at the time of such termination, shall supersede this definition with respect to that Participant. The determination of the Administrator as to the existence of Cause will be conclusive on the Participant and the Company.

11.7 **“Change in Control”** means (a) a sale of all or substantially all of the Company’s assets, or (b) any merger, amalgamation, consolidation or other business combination transaction of the Company with or into another corporation, entity or person, other than a transaction in which the holders of at least a majority of the voting shares of the Company outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into voting shares of the surviving entity) a majority of the total voting power represented by the voting shares of the Company (or the surviving entity) outstanding immediately after such transaction, or (c) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of the Company. Notwithstanding the foregoing, a Change in Control shall not be deemed to occur: (A) on account of the acquisition of voting shares by any institutional investor or any affiliate thereof or any other person, or persons acting as a group, that acquires the Company’s voting shares in a transaction or series of related transactions that are primarily a private financing transaction for the Company or (B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the **“Subject Person”**), exceeds the designated percentage threshold of the outstanding voting shares as a result of a repurchase or other acquisition of voting shares by the Company reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operating of this sentence) as a result of the acquisition of voting shares by the Company, and after such share acquisition, the Subject Person becomes the owner of any additional voting shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

Notwithstanding the foregoing, if a Change in Control constitutes a payment event with respect to any Award (or portion of any Award) that provides for the deferral of compensation that is subject to Section 409A, to the extent required to avoid the imposition of additional taxes under Section 409A, the transaction or event described in subsection (a), (b) or (c) with respect to such Award (or portion thereof) shall only constitute a Change in Control for purposes of the payment timing of such Award if such transaction also constitutes a “change in control event,” as defined in Treasury Regulation Section 1.409A-3(i)(5).

The Administrator shall have full and final authority, which shall be exercised in its discretion, to determine conclusively whether a Change in Control has occurred pursuant to the above definition, the date of the occurrence of such Change in Control and any incidental matters relating thereto; provided that any exercise of authority in conjunction with a determination of whether a Change in Control is a “change in control event” as defined in Treasury Regulation Section 1.409A-3(i)(5) shall be consistent with such regulation.

11.8 **“Code”** means the Internal Revenue Code of 1986, as amended, and the regulations issued thereunder.

11.9 **“Committee”** means one or more committees or subcommittees of the Board, which may include one or more Company directors or executive officers, to the extent Applicable Laws permit. To the extent required to comply with the provisions of Rule 16b-3, it is intended that each member of the Committee will be, at the time the Committee takes any action with respect to an Award that is subject to

Rule 16b-3, a “non-employee director” within the meaning of Rule 16b-3; however, a Committee member’s failure to qualify as a “non-employee director” within the meaning of Rule 16b-3 will not invalidate any Award granted by the Committee that is otherwise validly granted under the Plan.

11.10 “**Common Shares**” means the Class A Common Shares of the Company of par value US\$0.0001 each.

11.11 “**Company**” means Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company, or any successor.

11.12 “**Consultant**” means any person, including any adviser, engaged by the Company or its parent or Subsidiary to render services to such entity if the consultant or adviser: (i) renders bona fide services to the Company; (ii) renders services not in connection with the offer or sale of securities in a capital-raising transaction and does not directly or indirectly promote or maintain a market for the Company’s securities; and (iii) is a natural person.

11.13 “**Designated Beneficiary**” means the beneficiary or beneficiaries the Participant designates, in a manner the Administrator determines, to receive amounts due or exercise the Participant’s rights if the Participant dies or becomes incapacitated. Without a Participant’s effective designation, “Designated Beneficiary” will mean the Participant’s estate.

11.14 “**Director**” means a Board member.

11.15 “**Disability**” means a permanent and total disability under Section 22(e)(3) of the Code, as amended.

11.16 “**Dividend Equivalents**” means a right granted to a Participant under the Plan to receive the equivalent value (in cash or Shares) of dividends paid on Shares.

11.17 “**Employee**” means any employee of the Company or its Subsidiaries.

11.18 “**Equity Restructuring**” means a nonreciprocal transaction between the Company and its shareholders, such as a share dividend, bonus issue, share split, spin-off or recapitalization through a large, nonrecurring cash dividend, that affects the number or kind of Shares (or other Company securities) or the share price of Common Shares (or other Company securities) and causes a change in the per share value of the Common Shares underlying outstanding Awards.

11.19 “**Exchange Act**” means the Securities Exchange Act of 1934, as amended.

11.20 “**Fair Market Value**” means, as of any date, the value of Common Shares determined as follows: (i) if the Common Shares are listed on any established stock exchange, a share’s Fair Market Value will be the closing sales price for such Common Shares as quoted on such exchange for such date, or if no sale occurred on such date, the last day preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; (ii) if the Common Shares are not traded on a stock exchange but is quoted on a national market or other quotation system, the closing sales price on such date, or if no sales occurred on such date, then on the last date preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; or (iii) without an established market for the Common Shares, the Administrator will determine the Fair Market Value in its discretion. Notwithstanding the foregoing, with respect to any Award granted on the pricing date of the Company’s initial public offering, the Fair Market Value shall

mean the initial public offering price of a Share as set forth in the Company's final prospectus relating to its initial public offering filed with the Securities and Exchange Commission.

11.21 "**Greater Than 10% Shareholder**" means an individual then owning (within the meaning of Section 424(d) of the Code) more than 10% of the total combined voting power of all classes of shares of the Company or its parent or subsidiary corporation, as defined in Section 424(e) and (f) of the Code, respectively.

11.22 "**Incentive Stock Option**" means an Option intended to qualify as an "incentive stock option" as defined in Section 422 of the Code.

11.23 "**Non-Qualified Stock Option**" means an Option not intended or not qualifying as an Incentive Stock Option.

11.24 "**Option**" means an option to purchase Shares.

11.25 "**Other Share or Cash Based Awards**" means cash awards, awards of Shares, and other awards valued wholly or partially by referring to, or are otherwise based on, Shares or other property.

11.26 "**Overall Share Limit**" means (i) 4,466,500 Common Shares, (ii) any Common Shares which are subject to Prior Plan Awards which become available for issuance under the Plan pursuant to Article IV and (iii) an annual increase on the first day of each calendar year beginning January 1, 2019 and ending on and including January 1, 2028, equal to the lesser of (A) 4% of the aggregate number of Shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (B) such smaller number of Shares as is determined by the Board.

11.27 "**Participant**" means a Service Provider who has been granted an Award.

11.28 "**Performance Criteria**" mean the criteria (and adjustments) that the Administrator may select for an Award to establish performance goals for a performance period, which may include the following: net earnings or losses (either before or after one or more of interest, taxes, depreciation, amortization, and non-cash equity-based compensation expense); gross or net sales or revenue or sales or revenue growth; net income (either before or after taxes) or adjusted net income; profits (including but not limited to gross profits, net profits, profit growth, net operation profit or economic profit), profit return ratios or operating margin; budget or operating earnings (either before or after taxes or before or after allocation of corporate overhead and bonus); cash flow (including operating cash flow and free cash flow or cash flow return on capital); return on assets; return on capital or invested capital; cost of capital; return on shareholders' equity; total shareholder return; return on sales; costs, reductions in costs and cost control measures; expenses; working capital; earnings or loss per share; adjusted earnings or loss per share; price per share or dividends per share (or appreciation in or maintenance of such price or dividends); regulatory achievements or compliance; implementation, completion or attainment of objectives relating to research, development, regulatory, commercial, or strategic milestones or developments; market share; economic value or economic value added models; division, group or corporate financial goals; customer satisfaction/growth; customer service; employee satisfaction; recruitment and maintenance of personnel; human resources management; supervision of litigation and other legal matters; mergers, acquisitions, and other strategic partnerships, licenses, and other transactions; financial ratios (including those measuring liquidity, activity, profitability or leverage); debt levels or reductions; sales-related goals; financing and other capital raising transactions; cash on hand; acquisition activity; investment sourcing activity; and marketing initiatives, any of which may be measured in absolute terms or as compared to any incremental increase or decrease. Such performance goals also may be based solely by reference to the Company's performance or the performance of a Subsidiary, division, business segment or business unit of the

Company or a Subsidiary, or based upon performance relative to performance of other companies or upon comparisons of any of the indicators of performance relative to performance of other companies. The Committee may provide for exclusion of the impact of an event or occurrence which the Committee determines should appropriately be excluded, including (a) restructurings, discontinued operations, extraordinary items, and other unusual, infrequently occurring or non-recurring charges or events, (b) asset write-downs, (c) litigation or claim judgments or settlements, (d) acquisitions or divestitures, (e) reorganization or change in the corporate structure or capital structure of the Company, (f) an event either not directly related to the operations of the Company, Subsidiary, division, business segment or business unit or not within the reasonable control of management, (g) foreign exchange gains and losses, (h) a change in the fiscal year of the Company, (i) the refinancing or repurchase of bank loans or debt securities, (j) unbudgeted capital expenditures, (k) the issuance or repurchase of equity securities and other changes in the number of issued and outstanding shares, (l) conversion of some or all of convertible securities to Common Shares, (m) any business interruption event (n) the cumulative effects of tax or accounting changes in accordance with U.S. generally accepted accounting principles, or (o) the effect of changes in other laws or regulatory rules affecting reported results.

11.29 “**Plan**” means this 2018 Incentive Award Plan.

11.30 “**Prior Plans**” means, collectively, the Kiniksa Pharmaceuticals, Ltd. 2015 Equity Incentive Plan and any prior equity incentive plans of the Company or its predecessor.

11.31 “**Prior Plan Award**” means an award outstanding under the Prior Plans as of the Plan’s effective date in Section 10.3.

11.32 “**Public Trading Date**” means the first date upon which the Common Shares are listed (or approved for listing) upon notice of issuance on any securities exchange or designated (or approved for designation) upon notice of issuance as a national market security on an interdealer quotation system, or, if earlier, the date on which the Company becomes a “publicly held corporation” for purposes of Treasury Regulation Section 1.162-27(c)(1).

11.33 “**Restricted Share**” means a Share awarded to a Participant under Article VI subject to certain vesting conditions and other restrictions.

11.34 “**Restricted Share Unit**” means an unfunded, unsecured right to receive, on the applicable settlement date, one Share or an amount in cash or other consideration determined by the Administrator to be of equal value as of such settlement date, subject to certain vesting conditions and other restrictions.

11.35 “**Rule 16b-3**” means Rule 16b-3 promulgated under the Exchange Act.

11.36 “**Section 409A**” means Section 409A of the Code and all regulations, guidance, compliance programs and other interpretative authority thereunder.

11.37 “**Securities Act**” means the Securities Act of 1933, as amended.

11.38 “**Service Provider**” means an Employee, Consultant or Director.

11.39 “**Shares**” means Common Shares.

11.40 “**Share Appreciation Right**” means a share appreciation right granted under Article V.

11.41 “**Subject Person**” has the meaning set forth in Section 11.7.

11.42 “**Subsidiary**” means any entity (other than the Company), whether domestic or foreign, in an unbroken chain of entities beginning with the Company if each of the entities other than the last entity in the unbroken chain beneficially owns, at the time of the determination, securities or interests representing at least 50% of the total combined voting power of all classes of securities or interests in one of the other entities in such chain.

11.43 “**Substitute Awards**” shall mean Awards granted or Shares issued by the Company in assumption of, or in substitution or exchange for, awards previously granted, or the right or obligation to make future awards, in each case by a company acquired by the Company or any Subsidiary or with which the Company or any Subsidiary combines.

11.44 “**Termination of Service**” means the date the Participant ceases to be a Service Provider.

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**KINIKSA PHARMACEUTICALS, LTD.  
2018 INCENTIVE AWARD PLAN**

**SHARE OPTION GRANT NOTICE**

Capitalized terms not specifically defined in this Share Option Grant Notice (the “**Grant Notice**”) have the meanings given to them in the 2018 Incentive Award Plan (as amended from time to time, the “**Plan**”) of Kiniksa Pharmaceuticals, Ltd. (the “**Company**”).

The Company has granted to the participant listed below (“**Participant**”) the share option described in this Grant Notice (the “**Option**”), subject to the terms and conditions of the Plan and the Share Option Agreement attached as **Exhibit A** (the “**Agreement**”), both of which are incorporated into this Grant Notice by reference.

**Participant:**

**Grant Date:**

**Exercise Price per Share:**

**Shares Subject to the Option:**

**Final Expiration Date:**

**Vesting Commencement Date:**

**Vesting Schedule:** [To be specified in individual award agreements]

**Type of Option** [Incentive Stock Option/Non-Qualified Stock Option]

By Participant’s signature below, Participant agrees to be bound by the terms of this Grant Notice, the Plan and the Agreement. Participant has reviewed the Plan, this Grant Notice and the Agreement in their entirety, has had an opportunity to obtain the advice of counsel prior to executing this Grant Notice and fully understands all provisions of the Plan, this Grant Notice and the Agreement. Participant hereby agrees to accept as binding, conclusive and final all decisions or interpretations of the Administrator upon any questions arising under the Plan, this Grant Notice or the Agreement.

**KINIKSA PHARMACEUTICALS, LTD.**

**PARTICIPANT**

By: \_\_\_\_\_

Name: \_\_\_\_\_ [Participant Name]

Title: \_\_\_\_\_

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**SHARE OPTION AGREEMENT**

Capitalized terms not specifically defined in this Agreement have the meanings specified in the Grant Notice or, if not defined in the Grant Notice, in the Plan.

**ARTICLE I.  
GENERAL**

1.1 Grant of Option. The Company has granted to Participant the Option effective as of the grant date set forth in the Grant Notice (the “**Grant Date**”).

1.2 Incorporation of Terms of Plan. The Option is subject to the terms and conditions set forth in this Agreement and the Plan, which is incorporated herein by reference. In the event of any inconsistency between the Plan and this Agreement, the terms of the Plan will control.

**ARTICLE II.  
PERIOD OF EXERCISABILITY**

2.1 Commencement of Exercisability. The Option will vest and become exercisable according to the vesting schedule in the Grant Notice (the “**Vesting Schedule**”), except that any fraction of a Share as to which the Option would be vested or exercisable will be accumulated and will vest and become exercisable only when a whole Share has accumulated. In addition, if a Change in Control occurs (i) any outstanding portion of the Option that is not assumed, continued converted, replaced or substituted with a substantially similar award by the Company or a successor entity or its parent or subsidiary in the Change in Control (an “**Assumption**”) will be accelerated and will become vested and exercisable in full as of immediately prior to the occurrence of the Change in Control, and (ii) following an Assumption, if Participant’s employment with the Surviving Entity is terminated by the Surviving Entity without Cause within 12 months following the Change in Control, any outstanding portion of the Option will be accelerated and will become vested and exercisable in full as of immediately prior to Participant’s employment termination. Notwithstanding anything in the Grant Notice, the Plan or this Agreement to the contrary, unless the Administrator otherwise determines, the Option will immediately expire and be forfeited as to any portion that is not vested and exercisable as of Participant’s Termination of Service for any reason.

2.2 Duration of Exercisability. The Vesting Schedule is cumulative. Any portion of the Option which vests and becomes exercisable will remain vested and exercisable until the Option expires. The Option will be forfeited immediately upon its expiration.

2.3 Expiration of Option. The Option may not be exercised to any extent by anyone after, and will expire on, the first of the following to occur:

- (a) The final expiration date in the Grant Notice;
  - (b) Except as the Administrator may otherwise approve, the expiration of three (3) months from the date of Participant’s Termination of Service, unless Participant’s Termination of Service is for Cause or by reason of Participant’s death or Disability;
  - (c) Except as the Administrator may otherwise approve, the expiration of one (1) year from the date of Participant’s Termination of Service by reason of Participant’s death or Disability; and
-

(d) Except as the Administrator may otherwise approve, Participant's Termination of Service for Cause.

**ARTICLE III.  
EXERCISE OF OPTION**

3.1 Person Eligible to Exercise. During Participant's lifetime, only Participant may exercise the Option. After Participant's death, any exercisable portion of the Option may, prior to the time the Option expires, be exercised by Participant's Designated Beneficiary according to the procedures in the Plan.

3.2 Partial Exercise. Any exercisable portion of the Option or the entire Option, if then wholly exercisable, may be exercised, in whole or in part, according to the procedures in the Plan at any time prior to the time the Option or portion thereof expires, except that the Option may only be exercised for whole Shares.

3.3 Tax Withholding.

(a) The Company has the right and option, but not the obligation, to treat Participant's failure to provide timely payment in accordance with the Plan of any withholding tax arising in connection with the Option as Participant's election to satisfy all or any portion of the withholding tax by requesting the Company repurchase Shares otherwise issuable under the Option limited to the number of Shares which have a Fair Market Value on the date of repurchase necessary to pay the aggregate amount of tax liability.

(b) Participant acknowledges that Participant is ultimately liable and responsible for all taxes owed in connection with the Option, regardless of any action the Company or any Subsidiary takes with respect to any tax withholding obligations that arise in connection with the Option. Neither the Company nor any Subsidiary makes any representation or undertaking regarding the treatment of any tax withholding in connection with the awarding, vesting or exercise of the Option or the subsequent sale of Shares. The Company and the Subsidiaries do not commit and are under no obligation to structure the Option to reduce or eliminate Participant's tax liability.

**ARTICLE IV.  
OTHER PROVISIONS**

4.1 Adjustments. Participant acknowledges that the Option is subject to adjustment, modification and termination in certain events as provided in this Agreement and the Plan.

4.2 Notices. Any notice to be given under the terms of this Agreement to the Company must be in writing and addressed to the Company in care of the Company's Secretary at the Company's principal office or the Secretary's then-current email address or facsimile number. Any notice to be given under the terms of this Agreement to Participant must be in writing and addressed to Participant (or, if Participant is then deceased, to the person entitled to exercise the Option) at Participant's last known mailing address, email address or facsimile number in the Company's personnel files. By a notice given pursuant to this Section, either party may designate a different address for notices to be given to that party. Any notice will be deemed duly given when actually received, when sent by email, when sent by certified mail (return receipt requested) and deposited with postage prepaid in a post office or branch post office regularly maintained by the United States Postal Service, when delivered by a nationally recognized express shipping company or upon receipt of a facsimile transmission confirmation.

4.3 Titles. Titles are provided herein for convenience only and are not to serve as a basis for interpretation or construction of this Agreement.

4.4 Conformity to Securities Laws. Participant acknowledges that the Plan, the Grant Notice and this Agreement are intended to conform to the extent necessary with all Applicable Laws and, to the extent Applicable Laws permit, will be deemed amended as necessary to conform to Applicable Laws.

4.5 Successors and Assigns. The Company may assign any of its rights under this Agreement to single or multiple assignees, and this Agreement will inure to the benefit of the successors and assigns of the Company. Subject to the restrictions on transfer set forth in the Plan, this Agreement will be binding upon and inure to the benefit of the heirs, legatees, legal representatives, successors and assigns of the parties hereto.

4.6 Limitations Applicable to Section 16 Persons. Notwithstanding any other provision of the Plan or this Agreement, if Participant is subject to Section 16 of the Exchange Act, the Plan, the Grant Notice, this Agreement and the Option will be subject to any additional limitations set forth in any applicable exemptive rule under Section 16 of the Exchange Act (including any amendment to Rule 16b-3) that are requirements for the application of such exemptive rule. To the extent Applicable Laws permit, this Agreement will be deemed amended as necessary to conform to such applicable exemptive rule.

4.7 Entire Agreement. The Plan, the Grant Notice and this Agreement (including any exhibit hereto) constitute the entire agreement of the parties and supersede in their entirety all prior undertakings and agreements of the Company and Participant with respect to the subject matter hereof.

4.8 Agreement Severable. In the event that any provision of the Grant Notice or this Agreement is held illegal or invalid, the provision will be severable from, and the illegality or invalidity of the provision will not be construed to have any effect on, the remaining provisions of the Grant Notice or this Agreement.

4.9 Limitation on Participant's Rights. Participation in the Plan confers no rights or interests other than as herein provided. This Agreement creates only a contractual obligation on the part of the Company as to amounts payable and may not be construed as creating a trust. Neither the Plan nor any underlying program, in and of itself, has any assets. Participant will have only the rights of a general unsecured creditor of the Company with respect to amounts credited and benefits payable, if any, with respect to the Option, and rights no greater than the right to receive the Shares as a general unsecured creditor with respect to the Option, as and when exercised pursuant to the terms hereof.

4.10 Not a Contract of Employment. Nothing in the Plan, the Grant Notice or this Agreement confers upon Participant any right to continue in the employ or service of the Company or any Subsidiary or interferes with or restricts in any way the rights of the Company and its Subsidiaries, which rights are hereby expressly reserved, to discharge or terminate the services of Participant at any time for any reason whatsoever, with or without Cause, except to the extent expressly provided otherwise in a written agreement between the Company or a Subsidiary and Participant.

4.11 Counterparts. The Grant Notice may be executed in one or more counterparts, including by way of any electronic signature, subject to Applicable Law, each of which will be deemed an original and all of which together will constitute one instrument.

4.12 Incentive Stock Options. If the Option is designated as an Incentive Stock Option:

(a) Participant acknowledges that to the extent the aggregate fair market value of shares (determined as of the time the option with respect to the shares is granted) with respect to which share options intended to qualify as “incentive stock options” under Section 422 of the Code, including the Option, are exercisable for the first time by Participant during any calendar year exceeds \$100,000 or if for any other reason such share options do not qualify or cease to qualify for treatment as “incentive stock options” under Section 422 of the Code, such share options (including the Option) will be treated as non-qualified share options. Participant further acknowledges that the rule set forth in the preceding sentence will be applied by taking the Option and other share options into account in the order in which they were granted, as determined under Section 422(d) of the Code. Participant acknowledges that amendments or modifications made to the Option pursuant to the Plan that would cause the Option to become a Non-Qualified Stock Option will not materially or adversely affect Participant’s rights under the Option, and that any such amendment or modification shall not require Participant’s consent. Participant also acknowledges that if the Option is exercised more than three (3) months after Participant’s Termination of Service as an Employee, other than by reason of death or disability, the Option will be taxed as a Non-Qualified Stock Option.

(b) Participant will give prompt written notice to the Company of any disposition or other transfer of any Shares acquired under this Agreement if such disposition or other transfer is made (a) within two (2) years from the Grant Date or (b) within one (1) year after the transfer of such Shares to Participant. Such notice will specify the date of such disposition or other transfer and the amount realized, in cash, other property, assumption of indebtedness or other consideration, by Participant in such disposition or other transfer.

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**KINIKSA PHARMACEUTICALS, LTD.  
2018 INCENTIVE AWARD PLAN**

**RESTRICTED SHARE GRANT NOTICE**

Capitalized terms not specifically defined in this Restricted Share Grant Notice (the “**Grant Notice**”) have the meanings given to them in the 2018 Incentive Award Plan (as amended from time to time, the “**Plan**”) of Kiniksa Pharmaceuticals, Ltd. (the “**Company**”).

The Company has granted to the participant listed below (“**Participant**”) the Restricted Shares described in this Grant Notice (the “**Restricted Shares**”), subject to the terms and conditions of the Plan and the Restricted Share Agreement attached as **Exhibit A** (the “**Agreement**”), both of which are incorporated into this Grant Notice by reference.

**Participant:**

**Grant Date:**

**Number of Restricted Shares:**

**Vesting Commencement Date:**

**Vesting Schedule:** [To be specified in individual award agreements]

By Participant’s signature below, Participant agrees to be bound by the terms of this Grant Notice, the Plan and the Agreement. Participant has reviewed the Plan, this Grant Notice and the Agreement in their entirety, has had an opportunity to obtain the advice of counsel prior to executing this Grant Notice and fully understands all provisions of the Plan, this Grant Notice and the Agreement. Participant hereby agrees to accept as binding, conclusive and final all decisions or interpretations of the Administrator upon any questions arising under the Plan, this Grant Notice or the Agreement.

**KINIKSA PHARMACEUTICALS, LTD.**

**PARTICIPANT**

By: \_\_\_\_\_

Name: \_\_\_\_\_ [Participant Name]

Title: \_\_\_\_\_



**RESTRICTED SHARE AGREEMENT**

Capitalized terms not specifically defined in this Agreement have the meanings specified in the Grant Notice or, if not defined in the Grant Notice, in the Plan.

**ARTICLE I.  
GENERAL**

1.1 Issuance of Restricted Shares. The Company will issue the Restricted Shares to the Participant effective as of the grant date set forth in the Grant Notice and will cause (a) a share certificate or certificates representing the Restricted Shares to be registered in Participant's name or (b) the Restricted Shares to be held in book-entry form. If a share certificate is issued, the certificate will be delivered to, and held in accordance with this Agreement by, the Company or its authorized representatives and will bear the restrictive legends required by this Agreement. If the Restricted Shares are held in book-entry form, then the book-entry will indicate that the Restricted Shares are subject to the restrictions of this Agreement.

1.2 Incorporation of Terms of Plan. The Restricted Shares are subject to the terms and conditions set forth in this Agreement and the Plan, which is incorporated herein by reference. In the event of any inconsistency between the Plan and this Agreement, the terms of the Plan will control.

**ARTICLE II.  
VESTING, FORFEITURE AND ESCROW**

2.1 Vesting. The Restricted Shares will become vested Shares (the "**Vested Shares**") according to the vesting schedule in the Grant Notice except that any fraction of a Share that would otherwise become a Vested Share will be accumulated and will become a Vested Share only when a whole Vested Share has accumulated.

2.2 Forfeiture. In the event of Participant's Termination of Service for any reason, Participant will immediately and automatically forfeit to the Company any Shares that are not Vested Shares (the "**Unvested Shares**") at the time of Participant's Termination of Service, except as otherwise determined by the Administrator or provided in a binding written agreement between Participant and the Company. Upon forfeiture of Unvested Shares, the forfeited shares will be cancelled and Participant will have no further rights with respect to the Unvested Shares.

2.3 Escrow.

(a) Unvested Shares in certificated form will be held by the Company or its authorized representatives until (i) they are forfeited, (ii) they become Vested Shares or (iii) this Agreement is no longer in effect. By accepting this Award, Participant appoints the Company and its authorized representatives as Participant's attorney(s)-in-fact to take all actions necessary to effect cancellation of forfeited Unvested Shares (and Retained Distributions (as defined below), if any, paid on such forfeited Unvested Shares) to the Company as may be required pursuant to the Plan or this Agreement and to execute such representations or other documents or assurances as the Company or such representatives deem necessary or advisable in connection with any such transfer. The Company, or its authorized representative, will not be liable for any good faith act or omission with respect to the holding in escrow or transfer of the Restricted Shares.

(b) All cash dividends and other distributions made or declared with respect to Unvested Shares ("**Retained Distributions**") will be held by the Company until the time (if ever) when

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the Unvested Shares to which such Retained Distributions relate become Vested Shares. The Company will establish a separate Retained Distribution bookkeeping account ("**Retained Distribution Account**") for each Unvested Share with respect to which Retained Distributions have been made or declared in cash and credit the Retained Distribution Account (without interest) on the date of payment with the amount of such cash made or declared with respect to the Unvested Share. Retained Distributions (including any Retained Distribution Account balance) will immediately and automatically be forfeited to the Company upon forfeiture of the Unvested Share with respect to which the Retained Distributions were paid or declared.

(c) As soon as reasonably practicable following the date on which an Unvested Share becomes a Vested Share, the Company will (i) cause the certificate (or a new certificate without the legend required by this Agreement, if Participant so requests) representing the Share to be delivered to Participant or, if the Share is held in book-entry form, cause the notations indicating the Share is subject to the restrictions of this Agreement to be removed and (ii) pay to Participant the Retained Distributions relating to the Share.

2.4 Rights as Shareholder. Except as otherwise provided in this Agreement or the Plan, upon issuance of the Restricted Shares by the Company, Participant will have all the rights of a shareholder with respect to the Restricted Shares, including the right to vote the Restricted Shares and to receive dividends or other distributions paid or made with respect to the Restricted Shares.

### **ARTICLE III. TAXATION AND TAX WITHHOLDING**

3.1 Representation. Participant represents to the Company that Participant has reviewed with Participant's own tax advisors the tax consequences of the Restricted Shares and the transactions contemplated by the Grant Notice and this Agreement. Participant is relying solely on such advisors and not on any statements or representations of the Company or any of its agents.

3.2 Section 83(b) Election. If Participant makes an election under Section 83(b) of the Code with respect to the Restricted Shares, Participant will deliver a copy of the election to the Company promptly after filing the election with the Internal Revenue Service.

3.3 Tax Withholding.

(a) The Company has the right and option, but not the obligation, to treat Participant's failure to provide timely payment in accordance with the Plan of any withholding tax arising in connection with the Restricted Shares as Participant's election to satisfy all or any portion of the withholding tax by requesting the Company repurchase Shares otherwise deliverable under the Award limited to the number of Shares which have a Fair Market Value on the date of repurchase necessary to pay the aggregate amount of tax liability.

(b) Participant acknowledges that Participant is ultimately liable and responsible for all taxes owed in connection with the Restricted Shares, regardless of any action the Company or any Subsidiary takes with respect to any tax withholding obligations that arise in connection with the Restricted Shares. Neither the Company nor any Subsidiary makes any representation or undertaking regarding the treatment of any tax withholding in connection with the awarding, vesting or payment of the Restricted Shares or the subsequent sale of the Restricted Shares. The Company and the Subsidiaries do not commit and are under no obligation to structure this Award to reduce or eliminate Participant's tax liability.

**ARTICLE IV.  
RESTRICTIVE LEGENDS AND TRANSFERABILITY**

4.1 Legends. Any certificate representing a Restricted Share will bear the following legend until the Restricted Share becomes a Vested Share:

THE SHARES REPRESENTED BY THIS CERTIFICATE ARE SUBJECT TO FORFEITURE IN FAVOR OF THE COMPANY AND MAY BE TRANSFERRED ONLY IN ACCORDANCE WITH THE TERMS OF A RESTRICTED SHARE AGREEMENT BETWEEN THE COMPANY AND THE SHAREHOLDER, A COPY OF WHICH IS ON FILE WITH THE SECRETARY OF THE COMPANY.

4.2 Transferability. The Restricted Shares and any Retained Distributions are subject to the restrictions on transfer in the Plan and may not be sold, assigned or transferred in any manner unless and until they become Vested Shares. Any attempted transfer or disposition of Unvested Shares or related Retained Distributions prior to the time the Unvested Shares become Vested Shares will be null and void. The Company will not be required to (a) transfer on its books any Restricted Share that has been sold or otherwise transferred in violation of this Agreement or (b) treat as owner of such Restricted Share or accord the right to vote or pay dividends to any purchaser or other transferee to whom such Restricted Share has been so transferred. The Company may issue appropriate "stop transfer" instructions to its transfer agent, if any, or make appropriate notations to the same effect in its records.

**ARTICLE V.  
OTHER PROVISIONS**

5.1 Adjustments. Participant acknowledges that the Restricted Shares are subject to adjustment, modification and termination in certain events as provided in this Agreement and the Plan.

5.2 Notices. Any notice to be given under the terms of this Agreement to the Company must be in writing and addressed to the Company in care of the Company's Secretary at the Company's principal office or the Secretary's then-current email address or facsimile number. Any notice to be given under the terms of this Agreement to Participant must be in writing and addressed to Participant at Participant's last known mailing address, email address or facsimile number in the Company's personnel files. By a notice given pursuant to this Section, either party may designate a different address for notices to be given to that party. Any notice will be deemed duly given when actually received, when sent by email, when sent by certified mail (return receipt requested) and deposited with postage prepaid in a post office or branch post office regularly maintained by the United States Postal Service, when delivered by a nationally recognized express shipping company or upon receipt of a facsimile transmission confirmation.

5.3 Titles. Titles are provided herein for convenience only and are not to serve as a basis for interpretation or construction of this Agreement.

5.4 Conformity to Securities Laws. Participant acknowledges that the Plan, the Grant Notice and this Agreement are intended to conform to the extent necessary with all Applicable Laws and, to the extent Applicable Laws permit, will be deemed amended as necessary to conform to Applicable Laws.

5.5 Successors and Assigns. The Company may assign any of its rights under this Agreement to single or multiple assignees, and this Agreement will inure to the benefit of the successors and assigns of the Company. Subject to the restrictions on transfer set forth in this Agreement or the

Plan, this Agreement will be binding upon and inure to the benefit of the heirs, legatees, legal representatives, successors and assigns of the parties hereto.

5.6 Limitations Applicable to Section 16 Persons. Notwithstanding any other provision of the Plan or this Agreement, if Participant is subject to Section 16 of the Exchange Act, the Plan, the Grant Notice, this Agreement and the Restricted Shares will be subject to any additional limitations set forth in any applicable exemptive rule under Section 16 of the Exchange Act (including any amendment to Rule 16b-3) that are requirements for the application of such exemptive rule. To the extent Applicable Laws permit, this Agreement will be deemed amended as necessary to conform to such applicable exemptive rule.

5.7 Entire Agreement. The Plan, the Grant Notice and this Agreement (including any exhibit hereto) constitute the entire agreement of the parties and supersede in their entirety all prior undertakings and agreements of the Company and Participant with respect to the subject matter hereof.

5.8 Agreement Severable. In the event that any provision of the Grant Notice or this Agreement is held illegal or invalid, the provision will be severable from, and the illegality or invalidity of the provision will not be construed to have any effect on, the remaining provisions of the Grant Notice or this Agreement.

5.9 Limitation on Participant's Rights. Participation in the Plan confers no rights or interests other than as herein provided. This Agreement creates only a contractual obligation on the part of the Company as to amounts payable and may not be construed as creating a trust. Neither the Plan nor any underlying program, in and of itself, has any assets. Participant will have only the rights of a general unsecured creditor of the Company with respect to amounts credited and benefits payable, if any, with respect to the Award.

5.10 Not a Contract of Employment. Nothing in the Plan, the Grant Notice or this Agreement confers upon Participant any right to continue in the employ or service of the Company or any Subsidiary or interferes with or restricts in any way the rights of the Company and its Subsidiaries, which rights are hereby expressly reserved, to discharge or terminate the services of Participant at any time for any reason whatsoever, with or without cause, except to the extent expressly provided otherwise in a written agreement between the Company or a Subsidiary and Participant.

5.11 Counterparts. The Grant Notice may be executed in one or more counterparts, including by way of any electronic signature, subject to Applicable Law, each of which will be deemed an original and all of which together will constitute one instrument.

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**KINIKSA PHARMACEUTICALS, LTD.  
2018 INCENTIVE AWARD PLAN**

**RESTRICTED SHARE UNIT GRANT NOTICE**

Capitalized terms not specifically defined in this Restricted Share Unit Grant Notice (the “**Grant Notice**”) have the meanings given to them in the 2018 Incentive Award Plan (as amended from time to time, the “**Plan**”) of Kiniksa Pharmaceuticals, Ltd. (the “**Company**”).

The Company has granted to the participant listed below (“**Participant**”) the Restricted Share Units described in this Grant Notice (the “**RSUs**”), subject to the terms and conditions of the Plan and the Restricted Share Unit Agreement attached as **Exhibit A** (the “**Agreement**”), both of which are incorporated into this Grant Notice by reference.

**Participant:**

**Grant Date:**

**Number of RSUs:**

**Vesting Commencement Date:**

**Vesting Schedule:** [To be specified in individual award agreements]

By Participant’s signature below, Participant agrees to be bound by the terms of this Grant Notice, the Plan and the Agreement. Participant has reviewed the Plan, this Grant Notice and the Agreement in their entirety, has had an opportunity to obtain the advice of counsel prior to executing this Grant Notice and fully understands all provisions of the Plan, this Grant Notice and the Agreement. Participant hereby agrees to accept as binding, conclusive and final all decisions or interpretations of the Administrator upon any questions arising under the Plan, this Grant Notice or the Agreement.

**KINIKSA PHARMACEUTICALS, LTD.**

**PARTICIPANT**

By: \_\_\_\_\_

Name: \_\_\_\_\_ [Participant Name]

Title: \_\_\_\_\_



**RESTRICTED SHARE UNIT AGREEMENT**

Capitalized terms not specifically defined in this Agreement have the meanings specified in the Grant Notice or, if not defined in the Grant Notice, in the Plan.

**ARTICLE I.  
GENERAL**

1.1 Award of RSUs and Dividend Equivalents.

(a) The Company has granted the RSUs to Participant effective as of the grant date set forth in the Grant Notice (the "**Grant Date**"). Each RSU represents the right to receive one Share or, at the option of the Company, an amount of cash, in either case, as set forth in this Agreement. Participant will have no right to the distribution of any Shares or payment of any cash until the time (if ever) the RSUs have vested.

(b) The Company hereby grants to Participant, with respect to each RSU, a Dividend Equivalent for ordinary cash dividends paid to substantially all holders of outstanding Shares with a record date after the Grant Date and prior to the date the applicable RSU is settled, forfeited or otherwise expires. Each Dividend Equivalent entitles Participant to receive the equivalent value of any such ordinary cash dividends paid on a single Share. The Company will establish a separate Dividend Equivalent bookkeeping account (a "**Dividend Equivalent Account**") for each Dividend Equivalent and credit the Dividend Equivalent Account (without interest) on the applicable dividend payment date with the amount of any such cash paid.

1.2 Incorporation of Terms of Plan. The RSUs are subject to the terms and conditions set forth in this Agreement and the Plan, which is incorporated herein by reference. In the event of any inconsistency between the Plan and this Agreement, the terms of the Plan will control.

1.3 Unsecured Promise. The RSUs and Dividend Equivalents will at all times prior to settlement represent an unsecured Company obligation payable only from the Company's general assets.

**ARTICLE II.  
VESTING; FORFEITURE AND SETTLEMENT**

2.1 Vesting; Forfeiture. The RSUs will vest according to the vesting schedule in the Grant Notice except that any fraction of an RSU that would otherwise be vested will be accumulated and will vest only when a whole RSU has accumulated. In the event of Participant's Termination of Service for any reason, all unvested RSUs will immediately and automatically be cancelled and forfeited, except as otherwise determined by the Administrator or provided in a binding written agreement between Participant and the Company. Dividend Equivalents (including any Dividend Equivalent Account balance) will vest or be forfeited, as applicable, upon the vesting or forfeiture of the RSU with respect to which the Dividend Equivalent (including the Dividend Equivalent Account) relates.

2.2 Settlement.

(a) RSUs and Dividend Equivalents (including any Dividend Equivalent Account balance) will be paid in Shares or cash at the Company's option as soon as administratively practicable after the vesting of the applicable RSU, but in no event more than sixty (60) days after the RSU's vesting date. Notwithstanding the foregoing, the Company may delay any payment under this Agreement that the Company reasonably determines would violate Applicable Law until the earliest date the Company

reasonably determines the making of the payment will not cause such a violation (in accordance with Treasury Regulation Section 1.409A-2(b)(7)(ii)), provided the Company reasonably believes the delay will not result in the imposition of excise taxes under Section 409A.

(b) If an RSU is paid in cash, the amount of cash paid with respect to the RSU will equal the Fair Market Value of a Share on the day immediately preceding the payment date. If a Dividend Equivalent is paid in Shares, the number of Shares paid with respect to the Dividend Equivalent will equal the quotient, rounded down to the nearest whole Share, of the Dividend Equivalent Account balance divided by the Fair Market Value of a Share on the day immediately preceding the payment date.

### **ARTICLE III. TAXATION AND TAX WITHHOLDING**

3.1 Representation. Participant represents to the Company that Participant has reviewed with Participant's own tax advisors the tax consequences of this Award and the transactions contemplated by the Grant Notice and this Agreement. Participant is relying solely on such advisors and not on any statements or representations of the Company or any of its agents.

3.2 Tax Withholding.

(a) The Company has the right and option, but not the obligation, to treat Participant's failure to provide timely payment in accordance with the Plan of any withholding tax arising in connection with the RSUs or Dividend Equivalents as Participant's election to satisfy all or any portion of the withholding tax by requesting the Company repurchase Shares otherwise issuable under the Award limited to the number of Shares which have a Fair Market Value on the date of repurchase necessary to pay the aggregate amount of tax liability.

(b) Participant acknowledges that Participant is ultimately liable and responsible for all taxes owed in connection with the RSUs and the Dividend Equivalents, regardless of any action the Company or any Subsidiary takes with respect to any tax withholding obligations that arise in connection with the RSUs or Dividend Equivalents. Neither the Company nor any Subsidiary makes any representation or undertaking regarding the treatment of any tax withholding in connection with the awarding, vesting or payment of the RSUs or the Dividend Equivalents or the subsequent sale of Shares. The Company and the Subsidiaries do not commit and are under no obligation to structure the RSUs or Dividend Equivalents to reduce or eliminate Participant's tax liability.

### **ARTICLE IV. OTHER PROVISIONS**

4.1 Adjustments. Participant acknowledges that the RSUs, the Shares subject to the RSUs and the Dividend Equivalents are subject to adjustment, modification and termination in certain events as provided in this Agreement and the Plan.

4.2 Notices. Any notice to be given under the terms of this Agreement to the Company must be in writing and addressed to the Company in care of the Company's Secretary at the Company's principal office or the Secretary's then-current email address or facsimile number. Any notice to be given under the terms of this Agreement to Participant must be in writing and addressed to Participant at Participant's last known mailing address, email address or facsimile number in the Company's personnel files. By a notice given pursuant to this Section, either party may designate a different address for notices to be given to that party. Any notice will be deemed duly given when actually received, when sent by email, when sent by certified mail (return receipt requested) and deposited with postage prepaid in a post

office or branch post office regularly maintained by the United States Postal Service, when delivered by a nationally recognized express shipping company or upon receipt of a facsimile transmission confirmation.

4.3 Titles. Titles are provided herein for convenience only and are not to serve as a basis for interpretation or construction of this Agreement.

4.4 Conformity to Securities Laws. Participant acknowledges that the Plan, the Grant Notice and this Agreement are intended to conform to the extent necessary with all Applicable Laws and, to the extent Applicable Laws permit, will be deemed amended as necessary to conform to Applicable Laws.

4.5 Successors and Assigns. The Company may assign any of its rights under this Agreement to single or multiple assignees, and this Agreement will inure to the benefit of the successors and assigns of the Company. Subject to the restrictions on transfer set forth in the Plan, this Agreement will be binding upon and inure to the benefit of the heirs, legatees, legal representatives, successors and assigns of the parties hereto.

4.6 Limitations Applicable to Section 16 Persons. Notwithstanding any other provision of the Plan or this Agreement, if Participant is subject to Section 16 of the Exchange Act, the Plan, the Grant Notice, this Agreement, the RSUs and the Dividend Equivalents will be subject to any additional limitations set forth in any applicable exemptive rule under Section 16 of the Exchange Act (including any amendment to Rule 16b-3) that are requirements for the application of such exemptive rule. To the extent Applicable Laws permit, this Agreement will be deemed amended as necessary to conform to such applicable exemptive rule.

4.7 Entire Agreement. The Plan, the Grant Notice and this Agreement (including any exhibit hereto) constitute the entire agreement of the parties and supersede in their entirety all prior undertakings and agreements of the Company and Participant with respect to the subject matter hereof.

4.8 Agreement Severable. In the event that any provision of the Grant Notice or this Agreement is held illegal or invalid, the provision will be severable from, and the illegality or invalidity of the provision will not be construed to have any effect on, the remaining provisions of the Grant Notice or this Agreement.

4.9 Limitation on Participant's Rights. Participation in the Plan confers no rights or interests other than as herein provided. This Agreement creates only a contractual obligation on the part of the Company as to amounts payable and may not be construed as creating a trust. Neither the Plan nor any underlying program, in and of itself, has any assets. Participant will have only the rights of a general unsecured creditor of the Company with respect to amounts credited and benefits payable, if any, with respect to the RSUs and Dividend Equivalents, and rights no greater than the right to receive cash or the Shares as a general unsecured creditor with respect to the RSUs and Dividend Equivalents, as and when settled pursuant to the terms of this Agreement.

4.10 Not a Contract of Employment. Nothing in the Plan, the Grant Notice or this Agreement confers upon Participant any right to continue in the employ or service of the Company or any Subsidiary or interferes with or restricts in any way the rights of the Company and its Subsidiaries, which rights are hereby expressly reserved, to discharge or terminate the services of Participant at any time for any reason whatsoever, with or without Cause, except to the extent expressly provided otherwise in a written agreement between the Company or a Subsidiary and Participant.

4.11 Counterparts. The Grant Notice may be executed in one or more counterparts, including by way of any electronic signature, subject to Applicable Law, each of which will be deemed an original and all of which together will constitute one instrument.

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## 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 4, 2020

/s/ Sanj K. Patel

Sanj K. Patel

Chief Executive Officer and Chairman of the Board of Directors  
(Principal Executive Officer)

## CERTIFICATIONS

I, Michael R. Megna, 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 4, 2020

/s/ Michael R. Megna

Michael R. Megna  
VP, Finance and Chief Accounting Officer  
(Principal Financial Officer)

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**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, 2020 (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 4, 2020

/s/ Sanj K. Patel

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

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

I, Michael R. Megna, Chief Accounting 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, 2020 (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 4, 2020

/s/ Michael R. Megna

Michael R. Megna

VP, Finance and Chief Accounting Officer

(Principal Financial Officer)

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