Table of Contents

UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

FORM 10-Q

(Mark One)

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

For the quarterly period ended September 30, 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	**98-1327726**
(State or Other Jurisdiction of Incorporation or Organization)	(I.R.S. Employer Identification No.)

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

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

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

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

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

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

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

Large Accelerated Filer	☐	Accelerated Filer	☒
Non-accelerated Filer	☐	Smaller Reporting Company	☒
		Emerging Growth Company	☒

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☒

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

As of October 31, 2020, there were 68,075,035 common shares outstanding in aggregate, comprised of:
31,637,433 Class A common shares, par value $0.000273235 per share
2,355,458 Class B common shares, par value $0.000273235 per share
18,024,526 Class A1 common shares, par value $0.000273235 per share
16,057,618 Class B1 common shares, par value $0.000273235 per share

Kiniksa Pharmaceuticals, Ltd.

FORM 10-Q

FOR THE THREE AND NINE MONTHS ENDED SEPTEMBER 30, 2020

TABLE OF CONTENTS

	Page
PART I — FINANCIAL INFORMATION	5
Item 1. Financial Statements (unaudited)	5
Consolidated Balance Sheets as of September 30, 2020 and December 31, 2019	5
Consolidated Statements of Operations and Comprehensive Loss for the three and nine months ended September 30, 2020 and 2019	6
Consolidated Statements of Shareholders' Equity for the three and nine months ended September 30, 2020 and 2019	7
Consolidated Statements of Cash Flows for the nine months ended September 30, 2020 and 2019	8
Notes to Consolidated Financial Statements	9
Item 2. Management's Discussion and Analysis of Financial Condition and Results of Operations	32
Item 3. Quantitative and Qualitative Disclosures About Market Risk	47
Item 4. Controls and Procedures	47
PART II — OTHER INFORMATION	48
Item 1. Legal Proceedings	48
Item 1A. Risk Factors	48
Item 2. Unregistered Sales of Equity Securities and Use of Proceeds	122
Item 3. Defaults Upon Senior Securities	122
Item 4. Mine Safety Disclosures	122
Item 5. Other Information	122
Item 6. Exhibits	123
SIGNATURES	125

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 coronavirus disease 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;
- 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc. and Yescarta® is a registered trademark of Gilead Sciences, Inc., or its related companies.

Part I — Financial Information

Item 1. Financial Statements (unaudited)

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

	September 30, 2020	December 31, 2019
Assets		
Current assets:		
Cash and cash equivalents	$ 85,874	$ 46,928
Short-term investments	278,521	186,452
Prepaid expenses and other current assets	14,018	8,247
Total current assets	378,413	241,627
Property and equipment, net	4,573	6,398
Operating lease right-of-use assets	1,166	1,927
Restricted cash	210	210
Other long-term assets	48	—
Deferred tax assets	3	4,372
Total assets	$ 384,413	$ 254,534
Liabilities and Shareholders' Equity		
Current liabilities:		
Accounts payable	$ 3,925	$ 5,693
Accrued expenses	19,475	20,415
Operating lease liabilities	1,492	1,697
Other current liabilities	—	25
Total current liabilities	24,892	27,830
Non-current liabilities:		
Non-current operating lease liabilities	55	955
Other long-term liabilities	957	326
Total liabilities	25,904	29,111
Commitments and contingencies (Note 12)		
Shareholders' equity:		
Class A common shares, par value of $0.000273235 per share; 31,479,648 shares and 19,245,201 shares issued and outstanding as of September 30, 2020 and December 31, 2019, respectively	8	6
Class B common shares, par value of $0.000273235 per share; 2,355,458 shares and 4,638,855 shares issued and outstanding as of September 30, 2020 and December 31, 2019, respectively	1	1
Class A1 common shares, $0.000273235 par value; 18,024,526 shares and 14,995,954 issued and outstanding as of September 30, 2020 and December 31, 2019, respectively	5	4
Class B1 common shares, $0.000273235 par value; 16,057,618 shares issued and outstanding as of September 30, 2020 and December 31, 2019	4	4
Additional paid-in capital	822,301	581,467
Accumulated other comprehensive income	6	33
Accumulated deficit	(463,816)	(356,092)
Total shareholders' equity	358,509	225,423
Total liabilities and shareholders' equity	$ 384,413	$ 254,534

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 September 30,		Nine Months Ended September 30,	
	2020	2019	2020	2019
Operating expenses:				
Research and development	$ 31,419	$ 22,014	$ 74,644	$ 112,115
General and administrative	11,799	8,432	29,821	25,267
Total operating expenses	43,218	30,446	104,465	137,382
Loss from operations	(43,218)	(30,446)	(104,465)	(137,382)
Interest income	49	1,386	1,104	4,919
Loss before (provision) benefit for income taxes	(43,169)	(29,060)	(103,361)	(132,463)
(Provision) benefit for income taxes	(667)	2,002	(4,363)	2,393
Net loss	$ (43,836)	$ (27,058)	$ (107,724)	$ (130,070)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.66)	$ (0.49)	$ (1.80)	$ (2.42)
Weighted average common shares outstanding—basic and diluted	65,958,513	54,831,308	59,754,495	53,767,003
Comprehensive loss:				
Net loss	$ (43,836)	$ (27,058)	$ (107,724)	$ (130,070)
Other comprehensive income (loss):				
Unrealized gain (loss) on short-term investments and currency translation adjustments, net of tax	(3)	(39)	(27)	66
Total other comprehensive income	(3)	(39)	(27)	66
Total comprehensive loss	$ (43,839)	$ (27,097)	$ (107,751)	$ (130,004)

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				
Balances at December 31, 2019	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 and currency translation adjustments	—	—	—	207	—	207
Net loss	—	—	—	—	(26,419)	(26,419)
Balances at March 31, 2020	55,581,495	$ 15	$ 588,090	$ 240	$ (382,511)	$ 205,834
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 other offering costs	2,760,000	1	46,900	—	—	46,901
Issuance of Class A1 common shares upon completion of private placement, net of placement agent fees	1,600,000	—	27,594	—	—	27,594
Issuance of Class A common shares in connection with the release of escrow from the acquisition of all issued and outstanding equity securities of Primatope Therapeutics, Inc.	59,469	—	—	—	—	—
Exercise of options and issuance of shares under the employee share purchase plan	485,592	—	2,936	—	—	2,936
Share-based compensation expense	—	—	4,851	—	—	4,851
Unrealized loss on short-term investments and currency translation adjustments	—	—	—	(231)	—	(231)
Net loss	—	—	—	—	(37,469)	(37,469)
Balances at June 30, 2020	60,486,556	$ 16	$ 670,371	$ 9	$ (419,980)	$ 250,416
Issuance of Class A common shares upon completion of follow-on offering, net of underwriting discounts and commissions and other offering costs	5,952,381	2	117,691	—	—	117,693
Issuance of Class A1 common shares upon completion of private placement, net of placement agent fees	1,428,572	—	28,350	—	—	28,350
Issuance of Class A common shares in connection with the release of shares in connection with a milestone previously paid to Primatope Therapeutics, Inc.	16,634	—	—	—	—	—
Exercise of options	33,107	—	331	—	—	331
Share-based compensation expense	—	—	5,558	—	—	5,558
Unrealized loss on short-term investments and currency translation adjustments	—	—	—	(3)	—	(3)
Net loss	—	—	—	—	(43,836)	(43,836)
Balances at September 30, 2020	67,917,250	$ 18	$ 822,301	$ 6	$ (463,816)	$ 358,509

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income (Loss)	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
Balances at December 31, 2018	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 other offering costs	2,816,110	2	48,474	—	—	48,476
Issuance of Class A1 common shares upon completion of private placement, net of placement agent fees	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)
Balances at March 31, 2019	54,692,835	$ 15	$ 566,542	$ 8	$ (260,046)	$ 306,519
Class A common shares issued or to be issued in connection with a milestone payment due to Primatope Therapeutics, Inc.	94,284	—	1,800	—	—	1,800
Exercise of options and issuance of shares under the employee share purchase plan	70,573	—	512	—	—	512
Share-based compensation expense	—	—	3,464	—	—	3,464
Unrealized gain on short-term investments	—	—	—	93	—	93
Net loss	—	—	—	—	(37,191)	(37,191)
Balances at June 30, 2019	54,857,692	$ 15	$ 572,318	$ 101	$ (297,237)	$ 275,197
Exercise of options	13,648	—	53	—	—	53
Share-based compensation expense	—	—	3,758	—	—	3,758
Unrealized loss on short-term investments and currency translation adjustments	—	—	—	(39)	—	(39)
Net loss	—	—	—	—	(27,058)	(27,058)
Balances at September 30, 2019	54,871,340	$ 15	$ 576,129	$ 62	$ (324,295)	$ 251,911

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

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

	Nine Months Ended September 30,	
	2020	**2019**
Cash flows from operating activities:		
Net loss	$ (107,724)	$ (130,070)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	1,838	1,502
Share-based compensation expense	14,618	10,115
Class A common shares issued or to be issued as consideration for Primatope, including milestone payments	—	8,800
Loss on disposal of property and equipment	—	23
Non-cash lease expense	761	919
Accretion of discounts on short-term investments	(239)	(3,044)
Deferred income taxes	4,321	(3,411)
Changes in operating assets and liabilities:		
Prepaid expenses and other assets	(5,768)	(2,391)
Accounts payable	(1,704)	(4,504)
Accrued expenses and other liabilities	(969)	2,685
Accrued milestones	—	(14,850)
Operating lease liabilities	(1,105)	(916)
Other long-term liabilities	630	943
Net cash used in operating activities	(95,341)	(134,199)
Cash flows from investing activities:		
Purchases of property and equipment	(228)	(1,207)
Purchases of short-term investments	(344,159)	(406,135)
Proceeds from the maturities of short-term investments	252,300	454,640
Net cash provided by investing activities	(92,087)	47,298
Cash flows from financing activities:		
Proceeds from issuance of Class A common shares from follow-on offerings, net of underwriting discounts and commissions, inclusive of the over-allotment option exercise	165,725	48,595
Proceeds from issuance of Class A1 common shares from private placements, net of placement agent fees	55,944	34,511
Payments of offering costs	(976)	(118)
Proceeds from exercise of options and employee share purchase plan	5,681	746
Net cash provided by financing activities	226,374	83,734
Net increase in cash, cash equivalents and restricted cash	38,946	(3,167)
Cash, cash equivalents and restricted cash at beginning of period	47,138	72,186
Cash, cash equivalents and restricted cash at end of period	$ 86,084	$ 69,019
Supplemental information:		
Cash paid for income taxes	$ 482	$ 1,727
Supplemental disclosure of non-cash investing and financing activities:		
Deferred offering costs included in accrued expenses and accounts payable	$ 154	$ —
Property and equipment included in accrued expenses and accounts payable	$ 8	$ 371

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 coronavirus disease 2019 ("COVID-19") pandemic. 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. Upon approval from the U.S. Food and Drug Administration ("FDA"), if any, of a supplemental BLA ("sBLA") submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. The Company has never 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 ("CROs"), and contract manufacturing organizations ("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 or the easing of such measures, which continue to evolve. In December 2019, COVID-19 surfaced in Wuhan, China. The virus spread globally, and was declared a pandemic by the World Health Organization. The impact of this pandemic has been and will likely continue to be extensive on many aspects of society, which has resulted in and will likely continue to result in significant disruptions to healthcare systems, the global economy, as well as businesses and capital markets around the world.

In an effort to halt the spread of the COVID-19 pandemic, federal and state governments in the United States and the governments of other countries around the globe have implemented various measures in response to the pandemic, including significant restrictions on businesses and travel as well as social-distancing measures and the easing of such measures. 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 the Company's operations, it was and is considered to be an essential business so, to date, its operations have only been partially affected by these orders. In response to these orders, the Company implemented work-place rules and temporarily closed access to its California office space and restricted access to its Massachusetts facility to only those employees that needed to be in the office to execute their responsibilities and those employees who worked in the research and development laboratory space, with most of the employees continuing to carry out their responsibilities working outside of its offices. Subsequently in May and April 2020, the governors of Massachusetts and California, respectively, each announced a phased reopening plan for businesses and other organizations in their respective states. In response, the Company updated its work-place rules and designed a plan to reopen the Lexington and San Diego office spaces to additional groups of employees in phases, on an optional basis for now. Most of the employees continue to carry out their

responsibilities working outside of the Company's offices. As these measures implemented by federal and state governments in the United States in response to the pandemic continue to evolve, the Company continues to monitor the developments, restrictions and requirements in jurisdictions where it has offices, and plans to update the protocols for our offices as applicable.

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

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

Principles of Consolidation

The accompanying consolidated financial statements have been prepared in accordance with accounting principles generally accepted in the United States of America ("GAAP") and include the accounts of the Company and its wholly owned subsidiaries, Kiniksa Pharmaceuticals Corp. ("Kiniksa US"), Primatope Therapeutics, Inc. ("Primatope") and Kiniksa Pharmaceuticals (UK), Ltd. ("Kiniksa UK") as well as the subsidiaries of Kiniksa UK, Kiniksa Pharmaceuticals (Germany) GmbH ("Kiniksa Germany"), Kiniksa Pharmaceuticals (France) SARL ("Kiniksa France"), and Kiniksa Pharmaceuticals GmbH ("Kiniksa Switzerland"), after elimination of all significant intercompany accounts and transactions.

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 September 30, 2020 and during the three and nine 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.

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 September 30, 2020 and the results of its operations for the three and nine months ended September 30, 2020 and 2019 and its cash flows for the nine months ended September 30, 2020 and 2019. The results for the three and nine months ended September 30, 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, placement agent fees and other offering costs.

On May 18, 2020, the Company completed a follow-on offering of 2,760,000 Class A common shares, inclusive of the exercise of the underwriters’ overallotment option at a public offering price of $18.25 and a concurrent private placement of 1,600,000 Class A1 common shares at an offering price of $18.25 per share for aggregate gross proceeds of $79,570. The aggregate net proceeds to the Company from the follow-on offering and concurrent private

placement, inclusive of the over-allotment option exercise, was $74,495 after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

On July 24, 2020, the Company completed a follow-on offering of 5,952,381 Class A common shares, at a public offering price of $21.00 and a concurrent private placement of 1,428,572 Class A1 common shares at an offering price of $21.00 per share for aggregate gross proceeds of $155,000. The aggregate net proceeds to the Company from the follow-on offering and concurrent private placement was $146,043 after deducting underwriting discounts and commissions, placement agent fees 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 September 30, 2020, the Company had an accumulated deficit of $463,816. During the nine months ended September 30, 2020, the Company incurred a net loss of $107,724 and used $95,341 of cash in operating activities. The Company expects to continue to generate operating losses for the foreseeable future. As of September 30, 2020, the Company had cash, cash equivalents and short-term investments of $364,395.

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 September 30, 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 which are included in short-term investments on the Company's consolidated balance sheets are considered available-for-sale debt securities and are reported at fair value with unrealized gains and losses included as a component of shareholders' equity. Realized gains and losses, if any, on short-term investments are included in interest income.

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 its consolidated statements of cash flows to conform to current year presentation. The balances related to prepaid expenses and other assets and non-cash lease expense. The reclassifications had no effect on net cash used by operating activities or the Company's consolidated statements of operations and comprehensive loss.

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 September 30, 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 September 30, 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.

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

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 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.

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

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 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.

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 and nine months ended September 30, 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 and nine months ended September 30, 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, Kiniksa France, and Kiniksa Switzerland, are subject to taxation in their respective countries. Certain of the Company's subsidiaries, primarily Kiniksa US, operate under cost-plus arrangements.

The Company's U.S. provision for income taxes relates to current tax expense associated with the taxable income in the United States of its wholly owned subsidiary, Kiniksa US, as well as discrete tax events such as the

recognition of a valuation allowance. The current income tax expense 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. The Company has recorded an immaterial foreign provision for income taxes related to income in non-U.S. subsidiaries.

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

Recently Adopted Accounting Pronouncements

In 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.

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

3. Fair Value of Financial Assets and Liabilities

The following tables present information about the Company's financial instruments measured at fair value on a recurring basis and indicate the level of the fair value hierarchy used to determine such fair values:

	Fair Value Measurements as of September 30, 2020 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 210	$ —	$ —	$ 210
Cash equivalents — money market funds	31,043	—	—	31,043
Cash equivalents — U.S. Treasury notes	—	41,398	—	41,398
Short-term investments — U.S. Treasury notes	—	278,521	—	278,521
	$ 31,253	$ 319,919	$ —	$ 351,172

	Fair Value Measurements as of December 31, 2019 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
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
	$ 25,417	$ 196,644	$ —	$ 222,061

During the nine months ended September 30, 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 September 30, 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.

Short-term investments as of September 30, 2020 and December 31, 2019 consisted of U.S. Treasury notes which investments were each due within six months of such date.

	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Fair Value
September 30, 2020				
Short-term investments — U.S. Treasury notes	$ 278,514	$ 9	$ (2)	$ 278,521
	$ 278,514	$ 9	$ (2)	$ 278,521

	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Fair Value
December 31, 2019				
Short-term investments — U.S. Treasury notes	$ 186,415	$ 44	$ (7)	$ 186,452
	$ 186,415	$ 44	$ (7)	$ 186,452

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

As of September 30, 2020, the Company held eleven securities that were in an unrealized loss position. The aggregate fair value of securities held by the Company in an unrealized loss position was $78,056 at September 30, 2020. 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 was $43,107 at December 31, 2019. As of both September 30, 2020 and December 31, 2019, 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 September 30, 2020 and December 31, 2019.

4. Property and Equipment, Net

Property and equipment, net consisted of the following:

	September 30, 2020	December 31, 2019
Furniture, fixtures and vehicles	$ 47	$ 47
Computer hardware and software	349	344
Leasehold improvements	3,627	3,627
Lab equipment	4,713	4,685
Construction in progress	—	20
Total property and equipment	8,736	8,723
Less: Accumulated depreciation	(4,163)	(2,325)
Total property and equipment, net	$ 4,573	$ 6,398

Depreciation expense was $649 and $523 during the three months ended September 30, 2020 and 2019, respectively, and $1,838 and $1,502 during the nine months ended September 30, 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 January 31, 2021. On July 17, 2020, Kiniksa US entered into an amendment to extend the lease through January 31, 2022. Monthly lease payments for base rent are $13 and increase to $14 in accordance with the extension. Additional fees for ancillary charges such as the share of operating expenses, parking and real estate taxes are not included in the base rent.

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

The components of lease cost consisted of the following:

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2020	2019	2020	2019
Operating lease cost	$ 341	$ 340	$ 1,022	$ 1,102
Variable lease cost	30	52	132	161
Total lease cost	$ 371	$ 392	$ 1,154	$ 1,263

	September 30, 2020
Weighted-average remaining lease term (years)	0.90
Weighted-average discount rate	7.16%

Maturities of operating leases liabilities were as follows:

As of September 30,	
2020	$ 455
2021	1,124
2022	14
2023 and thereafter	—
Total future minimum lease payments	$ 1,593
Less imputed interest	(46)
Present value of lease liabilities	$ 1,547

6. Accrued Expenses

Accrued expenses consisted of the following:

	September 30, 2020	December 31, 2019
Accrued research and development expenses	$ 10,333	$ 11,813
Accrued employee compensation and benefits	6,747	7,089
Accrued legal and professional fees	1,621	1,087
Other	774	426
	$ 19,475	$ 20,415

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, placement agent fees and other offering costs.

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

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

On July 24, 2020, the Company completed a follow-on offering of 5,952,381 Class A common shares, at a public offering price of $21.00 and a concurrent private placement of 1,428,572 Class A1 common shares at an offering price of $21.00 per share for aggregate gross proceeds of $155,000. The aggregate net proceeds to the Company from the follow-on offering and concurrent private placement was $146,043 after deducting underwriting discounts and commissions, placement agent fees and other offering costs.

The rights of the holders of the Company's Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares are identical, except with respect to voting, transferability and conversion, as described below. As of September 30, 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.

Dividends

The common shareholders are entitled to receive dividends, as may be declared by the Company's board of directors. Through September 30, 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").

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

A total of 4,466,500 Class A common shares were initially reserved for issuance under the 2018 Plan. The number of Class A common shares that may be issued under the 2018 Plan will automatically increase on each January 1, beginning in 2019 and continuing for each fiscal year until, and including, the fiscal year ending December 31, 2028, equal to the lesser of (1) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (2) a smaller number of Class A common shares determined by the Company's board of directors. In December 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, lapses unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised, or forfeited under the 2018 Plan or the 2015 Plan will be added back to the Class A common shares available for issuance under the 2018 Plan. As of September 30, 2020, 1,610,263 shares remained available for future grant.

2015 Equity Incentive Plan

Until May 23, 2018 (the effective date of the 2018 Plan), the 2015 Plan provided for the Company to grant incentive share options, nonqualified share options, share grants and other share-based awards to employees and non-employees to purchase the Company's Class A common shares. On the effective date of the 2018 Plan, the Company ceased granting awards under the 2015 Plan. At that time, the 4,691,213 Class A common shares subject to outstanding awards under the 2015 Plan remained reserved for issuance under the plan pursuant to such awards and the 92,170 Class A common shares that had been available for future grant under the 2015 Plan were no longer authorized and reserved for issuance or available for future grant under the 2015 Plan.

As of September 30, 2020, there were 2,973,717 Class A common shares subject to outstanding awards under the 2015 Plan and reserved for issuance thereunder pursuant to such awards. The 2015 Plan continues to govern the terms and conditions of the outstanding awards granted under it. Class A common shares subject to awards granted under the 2015 Plan that expire, lapse unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised, or forfeited become available for issuance under the 2018 Plan.

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 (1) 1% of the Class A common

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

shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (2) a smaller number of Class A common shares determined by the Company's board of directors, provided that no more than 6,420,000 Class A common shares may be issued under the 2018 ESPP. In December 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. As of September 30, 2020, 549,300 Class A common shares were available for future issuance under the 2018 ESPP.

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	3,588,735	$ 11.42
Exercised	(1,113,532)	$ 3.13
Forfeited	(814,801)	$ 11.58
Outstanding as of September 30, 2020	10,152,136	$ 9.19
Options exercisable as of September 30, 2020	3,611,646	$ 8.20
Options unvested as of September 30, 2020	6,540,490	$ 10.46

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 and nine months ended September 30, 2020 and 2019 were as follows, presented on a weighted-average basis:

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2020	2019	2020	2019
Risk-free interest rate	0.36 %	1.69 %	0.56 %	2.13 %
Expected term (in years)	6.24	6.24	6.21	6.21
Expected volatility	82.58 %	80.37 %	81.02 %	79.24 %
Expected dividend yield	— %	— %	— %	— %

Rilonacept Long-Term Incentive Plan

In December 2019, the compensation committee of the Company's board of directors approved the Company's Rilonacept Long-Term Incentive Plan ("RLTIP") under the 2018 Plan to incentivize eligible employees of the Company or any of its subsidiaries to achieve FDA approval for the commercial sale and marketing of rilonacept for recurrent pericarditis in the United States ("RLTIP Milestone"). The RLTIP provides for the potential to receive a cash award and two grants of restricted share 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.

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

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 accordance with the RLTIP 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 RLTIP, 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 nine months ended September 30, 2020 and the year ended December 31, 2019, the Company also granted the First RSU Award as part of the RLTIP, 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 RLTIP Milestone is not achieved, the First RSU Award will not vest.

The grant-date fair value of the outstanding Time-Based RSUs remaining as of September 30, 2020 was $1,045 and is being recognized on a straight-line basis through the vest date for these RSUs. For the three and nine months ended September 30, 2020, the Company recognized $250 and $733 in Time-Based RSU expense, respectively. The grant-date fair value of the outstanding First RSU Award remaining as of September 30, 2020 was $2,762 and will be recognized when the RLTIP Milestone is deemed probable of achievement through the date the First RSU Award will vest. During the three and nine months ended September 30, 2020, the Company did not recognize compensation expense related to the First RSU Award, as achievement of the RLTIP Milestone was determined to be not probable.

The following table summarizes RSU activity for the nine months ended September 30, 2020:

	Number of Shares		Weighted Average Grant Date Fair Value
Unvested RSUs as of December 31, 2019	328,296	$	12.93
Granted	12,052	$	20.78
Vested	—	$	—
Forfeited	(52,837)	$	13.00
Unvested RSUs as of September 30, 2020	287,511	$	13.24

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

Share-Based Compensation

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

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2020	2019	2020	2019
Research and development expenses	$ 2,345	$ 1,435	$ 6,303	$ 4,126
General and administrative expenses	3,213	2,323	8,315	5,989
	$ 5,558	$ 3,758	$ 14,618	$ 10,115

9. License, Acquisition and Collaboration Agreements

Biogen Asset Purchase Agreement

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to vixarelimab and other antibodies covered by certain patent rights, including patents and other intellectual property rights, clinical data, know-how, and clinical drug supply. In addition, Biogen granted to the Company a non-exclusive, sublicensable, worldwide license to certain background patent rights related to the vixarelimab program. The Company is obligated to use commercially reasonable efforts to develop and commercialize such acquired products.

In exchange for these rights, the Company made an upfront payment to Biogen of $11,500 and a technology transfer payment of $500. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment and technology transfer payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

Under the Biogen Agreement, the Company is obligated to make milestone payments to Biogen of up to $179,000 upon the achievement of specified clinical and regulatory milestones in multiple indications in various territories, including milestone payments of $4,000 and $10,000 paid during the year ended December 31, 2017 and the year ended December 31, 2019, respectively, each payment was associated with the achievement of a specified clinical milestone event. No milestones were achieved or paid during the three and nine months ended September 30, 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 and nine months ended September 30, 2020, the Company recorded research and development expense of $14 and $92, respectively, primarily related to a milestone occurring in the first quarter of 2020 and the

annual maintenance fee both in connection with the retained contracts. During the three and nine months ended September 30, 2019, the Company recorded research and development expense of $164 and $10,330, respectively, primarily related to a milestone payment and other payments 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.

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, inclusive of its rights to develop and commercialize KPL-045, in accordance with the terms and conditions of the agreement.

The Company did not record any research and development expense during the three and nine months ended September 30, 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"). The aggregate amount of upfront and contingent payments the Company paid to the former Primatope shareholders to acquire the Company was comprised of (1) $15,000 paid at closing in March 2019, comprised of upfront consideration of $10,000 and milestone payments of $5,000, which had been achieved as of the closing date, and (2) $3,000 paid in June 2019 for the final milestone payment, which was achieved following the closing during the six months ended June 30, 2019, each paid in a

combination of cash and Class A common shares (inclusive of escrow and holdback shares 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 released the escrow and issued the shares that were held back at closing in June 2020 and issued the shares that were held back at the final milestone payment in September 2020. 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 and nine months ended September 30, 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 September 30, 2019, 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 nine months ended September 30, 2019, the Company recorded research and development expense of $18,000 related to the Primatope Acquisition.

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 and nine months ended September 30, 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, including, a $7,500 payment which may be met in the fourth quarter of 2020. Upon approval from the FDA, if any, of an sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, the Company will assume the sales and distribution of rilonacept for the approved indications in the United States and the Company would evenly split profits on sales of commercial products with Regeneron, after deducting certain commercialization expenses subject to specified limits.

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

The Company did not incur any research and development expense directly related to milestone payments due under the Regeneron Agreement during the three and nine months ended September 30, 2020 and 2019.

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 and nine months ended September 30, 2020, the Company did not incur any research and development expense related to the purchase of drug materials under this agreement. During the three and nine months ended September 30, 2019, the Company recorded research and development expense of $85 and $3,727, respectively, related to the purchase of drug materials under this agreement. As of September 30, 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 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 rilonacept 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.

MedImmune License Agreement

In December 2017, the Company entered into a license agreement (as amended from time to time, the "MedImmune Agreement") with MedImmune, Limited ("MedImmune"), pursuant to which MedImmune granted the Company an exclusive, sublicensable, worldwide license to certain intellectual property rights to make, use, develop and commercialize mavrilimumab. Under the MedImmune Agreement, the Company also acquired reference rights to relevant manufacturing and regulatory documents and MedImmune's existing supply of mavrilimumab drug substance and product. The Company is obligated to use commercially reasonable efforts to develop and commercialize the licensed products.

In exchange for these rights, the Company made an upfront payment of $8,000. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use. In addition, the Company is obligated to make clinical, regulatory and initial sales milestone payments of up to $72,500 in aggregate for the first two indications, including, a $5,000 pass-through payment due upon the achievement of a specified clinical milestone event which was 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. In July 2020, the Company entered

into an amendment to the MedImmune Agreement to establish a new coronavirus field and defer the payment of certain development and regulatory milestones as applied to the new coronavirus field. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

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

The MedImmune Agreement will expire upon the expiration of the royalty term in the last country for the last indication, as defined in the agreement. Either party may terminate the agreement upon the other party's insolvency or bankruptcy or for material breach of the agreement by the other party that remains uncured for 90 days. MedImmune has the right to terminate the agreement if the Company challenges any of the licensed patent rights. The Company may terminate the agreement at any time upon 90 days' prior written notice.

During the three and nine months ended September 30, 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 granulocyte-macrophage colony stimulating factor ("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 ("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. In August 2020, Kite as the sponsor of the study which had not begun to enroll, informed the Company that the clinical collaboration was being discontinued under a portfolio strategy review and terminated the agreement in accordance with its terms and conditions. During the three and nine months ended September 30, 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.

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

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

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2020	2019	2020	2019
Numerator:				
Net loss attributable to common shareholders	$ (43,836)	$ (27,058)	$ (107,724)	$ (130,070)
Denominator:				
Weighted average common shares outstanding—basic and diluted	65,958,513	54,831,308	59,754,495	53,767,003
Net loss per share attributable to common shareholders—basic and diluted	$ (0.66)	$ (0.49)	$ (1.80)	$ (2.42)

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 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 September 30,	
	2020	2019
Options to purchase common shares	10,152,136	8,635,968
Unvested RSUs	287,511	—
	10,439,647	8,635,968

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

The income tax rate for the three and nine months ended September 30, 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, U.S. federal and state research tax credits and the recognition of a valuation allowance against deferred tax assets. Income tax expense for the three and nine months ended September 30, 2020 was $667 and $4,363, respectively, and includes discrete tax expense primarily related to the recognition of a valuation allowance against deferred tax assets, partially offset by the tax benefits from share-based compensation taxable events.

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. As a result of significant cumulative tax benefits associated with share-based compensation taxable events recognized in the nine months ended September 30, 2020, the Company has significant negative evidence in the form of cumulative losses and believes that it is more likely than not that these United States deferred tax assets will not be utilized. As such, the Company recorded a valuation allowance against its U.S. deferred tax asset as of September 30, 2020. There are no material deferred tax assets in the other jurisdictions.

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 the Company with preclinical and clinical trial materials. As of September 30, 2020, the Company had committed to minimum payments under these agreements totaling $5,463.

Rilonacept Long-Term Incentive Plan

During the nine months ended September 30, 2020 and the year ended December 31, 2019, the Company granted a cash award and the First RSU Award to employees under the RLTIP. The cash award vests upon the achievement of the RLTIP Milestone, subject to the recipient’s continued employment through the date. The First RSU Award 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. As of September 30, 2020, the Company estimated cash payments of $1,990 under the RLTIP. In the event the RLTIP 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 September 30, 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 product candidates that are based on strong biologic rationale or validated mechanisms, target underserved conditions and offer the potential for differentiation. These product candidates are designed to modulate immunological pathways across a spectrum of diseases. Our product candidates include rilonacept, mavrilimumab, vixarelimab and KPL-404.

Rilonacept is an interleukin-1α, and interleukin-1β 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. In June 2020, we announced that our global, double-blind, placebo-controlled, randomized-withdrawal design, pivotal Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis, named RHAPSODY, met statistical significance on its primary and all major secondary efficacy endpoints, showing that rilonacept improved clinically meaningful outcomes associated with unmet medical need in recurrent pericarditis. Under the Regeneron Agreement, we are obligated to make payments to Regeneron of up to an aggregate of $27.5 million upon the achievement of specified regulatory milestones, including, a $7.5 million payment which may be met in the fourth quarter of 2020. If approved for recurrent pericarditis by the U.S. Food and Drug Administration, or FDA, we will commence the sales and distribution of rilonacept for the approved indications in the U.S. and evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. We continue to prepare for the potential commercial launch of rilonacept in recurrent pericarditis, which is anticipated in the first half of 2021, if approved by the FDA assuming priority review. In July 2020 we received Orphan Drug designation from the FDA for rilonacept for the treatment of pericarditis, which includes the treatment of recurrent pericarditis. We received Breakthrough Therapy designation from the FDA for rilonacept for the treatment of recurrent pericarditis in 2019.

Mavrilimumab is a monoclonal antibody that antagonizes granulocyte-macrophage colony stimulating factor, or GM-CSF. 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. We recently announced that the trial achieved both the primary and efficacy endpoint of time-to-first adjudicated GCA flare by Week 26 in all treated patients and the secondary efficacy endpoint of sustained remission at Week 26 in all treated patients with statistical significance. Additionally, while the trial was not powered for individual disease cohorts, there was a consistent trend of efficacy across the new onset and relapsing/refractory cohorts. In September 2020, the FDA granted Orphan Drug designation for mavrilimumab for the treatment of GCA. 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, for which Kite would be the sponsor and responsible for its conduct. In August 2020, Kite informed us that our clinical collaboration was being discontinued under a portfolio strategy review, which impacted the planned trial, and terminated the agreement in accordance with its terms. We are also evaluating mavrilimumab in severe coronavirus 2019 disease, or COVID-19, pneumonia and

hyperinflammation. In June 2020, we announced 28-day clinical outcomes from an open-label investigator-initiated treatment protocol with mavrilimumab conducted in Italy in 13 non-mechanically ventilated patients with severe COVID-19 pneumonia and hyperinflammation. Mavrilimumab-treated patients experienced earlier and improved clinical outcomes compared to control-group patients, including earlier weaning from supplemental oxygen, shorter hospitalizations, and no deaths. These data were published in *The Lancet Rheumatology*. We are enrolling the Phase 2 portion of a global, randomized, double-blind, placebo-controlled adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. Additionally, data from a randomized, double-blind, placebo-controlled investigator-initiated study in the same patient population in the United States are expected in the fourth quarter of 2020.

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 OSMRβ. We are evaluating vixarelimab for the potential treatment of with prurigo nodularis, a chronic inflammatory skin condition with an estimated U.S. prevalence of approximately 300,000 patients. In April 2020, we announced that our randomized, double-blind, placebo-controlled Phase 2a trial of vixarelimab in prurigo nodularis met its primary efficacy endpoint: there was a statistically significant reduction in weekly-average Worst-Itch Numeric Rating Scale (WI-NRS) from baseline at Week 8 in vixarelimab recipients compared to placebo recipients. Additionally, a statistically significant percentage of vixarelimab recipients achieved a prurigo nodularis-investigator's global assessment (PN-IGA) score of 0/1 at Week 8 compared to placebo recipients, and the majority of vixarelimab recipients showed a clinically meaningful greater-than-or-equal-to 4-point weekly-average WI-NRS reduction at Week 8. In May 2020, we announced data from an exploratory randomized, double-blind, placebo-controlled Phase 2 clinical trial in diseases characterized by chronic pruritus, including plaque psoriasis, chronic idiopathic pruritus, lichen simplex chronicus, chronic idiopathic urticaria and lichen planus. The plaque psoriasis cohort achieved a statistically significant reduction in weekly-average WI-NRS at Week 8. Additionally, the lichen simplex chronicus, chronic idiopathic urticaria and lichen planus cohorts showed encouraging efficacy results as measured by percent change from baseline in weekly-average WI-NRS at Week 8. We expect to initiate a dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis in the fourth quarter 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 to evaluate safety and pharmacokinetics well as receptor occupancy and T-cell dependent antibody response, or TDAR, in these subjects. The study is divided into two parts: single doses of KPL-404 0.03 mg/kg, 0.3 mg/kg, 1 mg/kg, 3 mg/kg or 10 mg/kg intravenous and single doses of KPL-404 1 mg/kg or 5 mg/kg subcutaneous. We expect pharmacokinetic, receptor occupancy and TDAR data from the first cohorts, including the 3 mg/kg IV dose level, in the fourth quarter of 2020. Further, we expect final data and safety follow-up from all cohorts in the first half of 2021.

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 announced positive data from the global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, named RHAPSODY. Upon approval from the FDA, if any, of the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. However, we have not yet demonstrated our ability to successfully 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, placement agent fees and other offering costs.

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

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

We have incurred significant operating losses since inception. Our ability to generate product revenue sufficient to achieve profitability will depend heavily on the successful development and eventual commercialization of one or more of our current or future product candidates. Our net losses were $107.7 million and $130.1 million for the nine months ended September 30, 2020 and 2019, respectively. As of September 30, 2020, we had an accumulated deficit of $463.8 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.

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 September 30, 2020, we had cash, cash equivalents and short-term investments of $364.4 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 September 30,		Nine Months Ended September 30,	
	2020	2019	2020	2019
	(in thousands)		(in thousands)	
Rilonacept	$ 4,812	$ 5,461	$ 13,981	$ 19,766
Mavrilimumab	11,925	2,808	17,469	11,512
Vixarelimab [1]	1,278	2,469	5,741	25,487
KPL-404 [2]	880	1,227	2,927	21,826
Unallocated research and development expenses	12,524	10,049	34,526	33,524
Total research and development expenses	$ 31,419	$ 22,014	$ 74,644	$ 112,115

(1) The amount for the nine months ended September 30, 2019 includes expense of $10.0 million related to an accrued 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 nine months ended September 30, 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 share 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.

We announced positive data from our global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, named RHAPSODY. Upon approval from the FDA, if any, of the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States. However, we have not yet demonstrated our ability to successfully obtain regulatory approvals, manufacture a commercial scale drug, or conduct sales and marketing activities. As such, the successful development and commercialization of rilonacept and our other 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 clinical development of rilonacept and any of our product candidates or when, if ever, material net cash inflows may commence from any of our product candidates, if approved. This uncertainty is due to the numerous risks and uncertainties associated with product development and commercialization, including the uncertainty of:

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

- the timing, receipt and terms of any marketing approvals from applicable regulatory authorities, including the FDA;
- the extent of any required post-marketing approval commitments to applicable regulatory authorities;
- increasing clinical and commercial manufacturing capabilities or making arrangements with additional third-party manufacturers to successfully manufacture our product candidates;
- development and timely delivery of clinical-grade and commercial-grade drug formulations that can be used in our clinical trials and for commercial launch;
- establishing a sales, marketing and distribution infrastructure to commercialize products for which we may obtain marketing approval;
- launching commercial sales of our product candidates, if and when approved, whether alone or in collaboration with others;
- making milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreements;
- obtaining, maintaining, defending and enforcing patent claims and other intellectual property rights;
- significant and changing government regulation; and
- maintaining a continued acceptable safety profile of 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, and that such costs will increase over time especially as we have determined that we will be a large accelerated filer and cease being an “emerging growth company” as of December 31, 2020 and cease being a “smaller reporting company” as of January 1, 2021. After we cease being an emerging growth company and smaller reporting company, we will no longer be permitted to rely on exemptions from certain requirements that are applicable to public companies that are not emerging growth companies or smaller reporting companies.

Interest Income

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

Income Taxes

As an exempted company incorporated under the laws of Bermuda, we are principally subject to taxation in Bermuda. Under the current laws of Bermuda, tax on a company’s income is assessed at a zero percent tax rate. As a

result, we have not recorded any income tax benefits from our losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards are currently available to us for those losses, while our assets remain in Bermuda. Our wholly owned U.S. subsidiaries, Kiniksa Pharmaceuticals Corp., or Kiniksa US, and Primatope are subject to federal and state income taxes in the United States. Our wholly owned subsidiary Kiniksa Pharmaceuticals (UK), Ltd., and its wholly owned subsidiaries, Kiniksa Pharmaceuticals (Germany) GmbH, Kiniksa Pharmaceuticals (France) SARL, and Kiniksa Pharmaceuticals GmbH are subject to taxation in their respective countries. Our provision for income taxes relates mainly to U.S. taxable income, generated by our wholly owned subsidiary Kiniksa US.

Results of Operations

Comparison of the Three Months Ended September 30, 2020 and 2019

The following table summarizes our results of operations for the three months ended September 30, 2020 and 2019:

	Three Months Ended September 30,		
	2020	2019	Change
	(in thousands)		
Operating expenses:			
Research and development	$ 31,419	$ 22,014	$ 9,405
General and administrative	11,799	8,432	3,367
Total operating expenses	43,218	30,446	12,772
Loss from operations	(43,218)	(30,446)	(12,772)
Interest income	49	1,386	(1,337)
Loss before (provision) benefit for income taxes	(43,169)	(29,060)	(14,109)
(Provision) benefit for income taxes	(667)	2,002	(2,669)
Net loss	$ (43,836)	$ (27,058)	$ (16,778)

Research and Development Expenses

	Three Months Ended September 30,		
	2020	2019	Change
	(in thousands)		
Direct research and development expenses by program:			
Rilonacept	$ 4,812	$ 5,461	$ (649)
Mavrilimumab	11,925	2,808	9,117
Vixarelimab	1,278	2,469	(1,191)
KPL-404	880	1,227	(347)
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	8,274	7,103	1,171
Other	4,250	2,946	1,304
Total research and development expenses	$ 31,419	$ 22,014	$ 9,405

Research and development expenses were $31.4 million for the three months ended September 30, 2020, compared to $22.0 million for the three months ended September 30, 2019, or an increase of $9.4 million.

During the three months ended September 30, 2020, the research and development expenses incurred related to external spend for our research programs of $18.9 million, including $8.0 million of preclinical and clinical trial costs, and $8.5 million of clinical drug supply related costs. In addition, we incurred unallocated research and development expenses of $12.5 million including, $8.3 million of personnel costs and $4.2 million of other operating costs including costs associated with our laboratory. During the three months ended September 30, 2019, research and development expenses incurred related to external spend for our research programs of $12.0 million, including $7.6 million of preclinical and clinical trial costs and $1.5 million of clinical drug supply related costs. In addition, we incurred

unallocated research and development expense of $10.0 million including $7.1 million of personnel costs and $2.9 million of other operating costs including costs associated with our laboratory.

The direct costs for our rilonacept program were $4.8 million during the three months ended September 30, 2020, compared to $5.5 million during the three months ended September 30, 2019, or a decrease of $0.7 million. During the three months ended September 30, 2020, expenses incurred primarily related to conducting RHAPSODY, our global, pivotal Phase 3 clinical trial in recurrent pericarditis, and supply chain costs, compared to the three months ended September 30, 2019, in which expenses incurred related to our clinical research and development for RHAPSODY.

The direct costs for our mavrilimumab program were $11.9 million during the three months ended September 30, 2020, compared to $2.8 million during the three months ended September 30, 2019, or an increase of $9.1 million. During the three months ended September 30, 2020, expenses incurred related primarily to our global Phase 2 clinical trial in GCA, including manufacturing costs for our clinical drug supply, and start-up costs for the phase 2 portion of our Phase 2/3 clinical trial in COVID-19 pneumonia and hyperinflammation, compared to the three months ended September 30, 2019, in which expenses incurred related primarily to our global Phase 2 clinical trial in GCA and manufacturing process development related expenses.

The direct costs for our vixarelimab program were $1.3 million during the three months ended September 30, 2020, compared to $2.5 million during the three months ended September 30, 2019, or a decrease of $1.2 million. During the three months ended September 30, 2020, expenses incurred primarily related to completing our Phase 2a clinical trial in prurigo nodularis and our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus. In the three months ended September 30, 2019, expenses incurred related to our Phase 2a clinical trial in prurigo nodularis and our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus, including manufacturing costs for our clinical drug supply.

The direct costs for our KPL-404 program were $0.9 million during the three months ended September 30, 2020, compared to $1.2 million during the three months ended September 30, 2019, or a decrease of $0.3 million. During the three months ended September 30, 2020, expenses incurred primarily related to limited clinical trial expenses for our Phase 1 trial of KPL-404 in healthy volunteers due to delays associated with the COVID-19 pandemic, including toxicology costs. During the three months ended September 30, 2019, expenses incurred primarily related to preclinical research and development, including manufacturing development costs.

Unallocated research and development expenses were $12.5 million for the three months ended September 30, 2020 compared to $10.0 million for the three months ended September 30, 2019. The increase of $2.5 million in unallocated research and development expenses was due to an increase of $1.2 million in personnel-related costs, primarily related to share-based compensation and an increase of $1.1 million in operating costs associated with our laboratory and general research costs, as compared to the three months ended September 30, 2019. Personnel-related costs for the three months ended September 30, 2020 and 2019 included share-based compensation of $2.3 million and $1.4 million, respectively.

General and Administrative Expenses

General and administrative expenses were $11.8 million for the three months ended September 30, 2020 compared to $8.4 million for the three months ended September 30, 2019. The increase of $3.4 million was primarily due to increases of $1.5 million in personnel-related costs and $1.4 million in costs associated with pre-commercialization activities of our rilonacept program. Personnel-related costs for the three months ended September 30, 2020 and 2019 included share-based compensation of $3.2 million and $2.3 million, respectively.

Interest Income

Interest income was $0.1 million for the three months ended September 30, 2020 compared to $1.4 million for the three months ended September 30, 2019. The decrease was due primarily to lower interest rates on U.S. Treasury notes.

Provision for Income Taxes

For the three months ended September 30, 2020, we recorded a provision for income taxes of $0.7 million relating primarily to the tax impact from the current tax expense due to income from our cost plus arrangement in the United States, net of R&D credits utilized offset by tax benefit related to the exercise of share options.

Comparison of the Nine Months Ended September 30, 2020 and 2019

The following table summarizes our results of operations for the nine months ended September 30, 2020 and 2019:

	Nine Months Ended September 30,		
	2020	2019	Change
		(in thousands)	
Operating expenses:			
Research and development	$ 74,644	$ 112,115	$ (37,471)
General and administrative	29,821	25,267	4,554
Total operating expenses	104,465	137,382	(32,917)
Loss from operations	(104,465)	(137,382)	32,917
Interest income	1,104	4,919	(3,815)
Loss before (provision) benefit for income taxes	(103,361)	(132,463)	29,102
(Provision) benefit for income taxes	(4,363)	2,393	(6,756)
Net loss	$ (107,724)	$ (130,070)	$ 22,346

Research and Development Expenses

	Nine Months Ended September 30,		
	2020	2019	Change
		(in thousands)	
Direct research and development expenses by program:			
Rilonacept	$ 13,981	$ 19,766	$ (5,785)
Mavrilimumab	17,469	11,512	5,957
Vixarelimab	5,741	25,487	(19,746)
KPL-404	2,927	21,826	(18,899)
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	24,246	21,834	2,412
Other	10,280	11,690	(1,410)
Total research and development expenses	$ 74,644	$ 112,115	$ (37,471)

Research and development expenses were $74.6 million for the nine months ended September 30, 2020, compared to $112.1 million for the nine months ended September 30, 2019, or a decrease of $37.5 million.

During the nine months ended September 30, 2020, the research and development expenses incurred related to external spend for our research programs of $40.1 million, including $22.5 million of preclinical and clinical trial costs and $11.5 million of clinical drug supply related costs. In addition, we incurred unallocated research and development expenses of $34.5 million, including $24.2 million of personnel costs and $10.3 million of other operating costs including costs associated with our laboratory. During the nine months ended September 30, 2019, expenses incurred related to the acquisition of all of the issued and outstanding equity securities of Primatope for aggregate upfront and contingent payments of $18.0 million, paid in a combination of cash and Class A common shares (inclusive of escrow and holdback share amounts) in accordance with the Primatope Agreement, $10.0 million for an accrued milestone under the Biogen Agreement, associated with the achievement of a specified clinical milestone event, and expenses for external spend for our research programs of $50.6 million, including $28.0 million of preclinical and clinical trial costs

and $14.7 million of clinical drug supply related costs. In addition, we incurred unallocated research and development expenses of $33.5 million, including, $21.9 million of personnel costs and $11.7 million of other operating costs including costs associated with our laboratory.

The direct costs for our rilonacept program were $14.0 million during the nine months ended September 30, 2020, compared to $19.8 million during the nine months ended September 30, 2019, or a decrease of $5.8 million. During the nine months ended September 30, 2020, expenses incurred primarily related to clinical trial costs for conducting RHAPSODY and supply chain costs. During the nine months ended September 30, 2019, expenses incurred primarily related to our clinical research and development for RHAPSODY, including $3.6 million related to purchases of drug materials under our clinical supply agreement with Regeneron, as well as for our open-label Phase 2 proof-of-concept clinical trial

The direct costs for our mavrilimumab program were $17.5 million during the nine months ended September 30, 2020, compared to $11.5 million during the nine months ended September 30, 2019, or an increase of $6.0 million. During the nine months ended September 30, 2020, expenses incurred related primarily to our global Phase 2 clinical trial in GCA, including manufacturing costs for our clinical drug supply, and start-up costs for the Phase 2 portion of our Phase 2/3 clinical trial in COVID-19 pneumonia and hyperinflammation, compared to the nine months ended September 30, 2019, in which expenses incurred related primarily clinical trial expenses related to our global Phase 2 trial in GCA.

The direct costs for our vixarelimab program were $5.7 million during the nine months ended September 30, 2020, compared to $25.5 million during the nine months ended September 30, 2019, or a decrease of $19.8 million. During the nine months ended September 30, 2020, expenses incurred primarily related to clinical trial costs for our Phase 2a clinical trial in prurigo nodularis and our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus, compared to the nine months ended September 30, 2019, in which expenses incurred related to a milestone payment of $10.0 million under the Biogen Agreement associated with the achievement of a specified clinical milestone event as well as expenses incurred for our Phase 2a clinical trial in prurigo nodularis, our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus and our Phase 1b clinical trial, as well as approximately $1.6 million of manufacturing and development costs for our clinical drug supply.

The direct costs for our KPL-404 program were $2.9 million during the nine months ended September 30, 2020, compared to $21.8 million during the nine months ended September 30, 2019, or a decrease of $18.9 million. During the nine months ended September 30, 2020, expenses incurred primarily related to preclinical and clinical trial costs for our Phase 1 trial of KPL-404 in healthy volunteers, including start-up and toxicology costs. During the nine months ended September 30, 2019, expenses incurred primarily related to the acquisition of all of the issued and outstanding equity securities of Primatope for aggregate upfront and contingent payments of $18.0 million paid in a combination of cash and Class A common shares (inclusive of escrow and holdback share amounts) in accordance with the Primatope Agreement.

Unallocated research and development expenses were $34.5 million for the nine months ended September 30, 2020 compared to $33.5 million for the nine months ended September 30, 2019. The increase of $1.0 million in unallocated research and development expenses was due to increases of $2.1 million in operating costs of our laboratory and $2.4 million in personnel-related costs primarily related to share-based compensation, partially offset by decreases of $2.0 million related to the cessation of our development of KPL-045 and $1.4 million in other operating expenses. Personnel-related costs for the nine months ended September 30, 2020 and 2019 included share-based compensation of $6.3 million and $4.1 million, respectively.

General and Administrative Expenses

General and administrative expenses were $29.8 million for the nine months ended September 30, 2020 compared to $25.3 million for the nine months ended September 30, 2019. The increase of $4.5 million was primarily due to increases of $3.3 million in personnel-related costs and $3.0 million in marketing costs associated with the pre-commercialization of our rilonacept program partially offset by a decrease $1.7 million of other expenses primarily due to a decrease of travel costs due to the travel restrictions associated with the COVID-19 pandemic and other

miscellaneous professional fees. Personnel-related costs for the nine months ended September 30, 2020 and 2019 included share-based compensation of $8.3 million and $6.0 million, respectively.

Interest Income

Interest income was $1.1 million for the nine months ended September 30, 2020 compared to $4.9 million for the nine months ended September 30, 2019. The decrease was primarily due to lower interest rates on U.S. Treasury notes.

Provision for Income Taxes

For the nine months ended September 30, 2020, we recorded a provision for income taxes of $4.4 million relating primarily to the tax impact from the recognition of a valuation allowance reserving against our existing deferred tax assets and current tax expense offset by tax benefit related to 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, placement agent fees and other offering costs.

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

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

As of September 30, 2020, we had cash, cash equivalents and short-term investments of $364.4 million.

Cash Flows

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

	Nine Months Ended September 30,	
	2020	2019
	(in thousands)	
Net cash used in operating activities	$ (95,341)	$ (134,199)
Net cash (used) provided by investing activities	(92,087)	47,298
Net cash provided by financing activities	226,374	83,734
Net increase in cash and cash equivalents and restricted cash	$ 38,946	$ (3,167)

Operating Activities

During the nine months ended September 30, 2020, operating activities used $95.3 million of cash, primarily resulting from our net loss of $107.7 million and net cash used in our operating assets and liabilities of $8.9 million, partially offset by non-cash charges of $21.3 million. Net cash used in our operating assets and liabilities for the nine months ended September 30, 2020 consisted of a $1.7 million decrease in accounts payable primarily due to the timing of vendor invoicing and payments, a $1.0 decrease in accrued expenses and other liabilities primarily due to decreases in the accrued costs for our research and manufacturing activities and the cash payment of the 2019 employee bonuses offset by increases in the accrued costs for our clinical trials and pre-commercialization activities of our rilonacept program, a $1.1 million decrease in operating lease liabilities due to monthly payments for our right-of-use assets, and a $5.8 million increase in prepaid expenses and other current assets due to increases in prepaid expenses to CROs related to our clinical trials.

During the nine months ended September 30, 2019, operating activities used $134.2 million of cash, primarily resulting from our net loss of $130.1 million, partially offset by non-cash charges of $14.9 million and net cash used by changes in our operating assets and liabilities of $19.0 million. Net cash used by changes in our operating assets and liabilities for the nine months ended September 30, 2019 consisted of a $14.9 million decrease in accrued milestones, a $4.5 million decrease in accounts payable, a $0.9 million decrease in operating lease liabilities, and a $2.4 million increase in prepaid expenses and other current assets, partially offset by a $0.9 million increase in other long-term liabilities and a $2.7 million increase in accrued expenses. The decrease in accrued milestones resulted from the payment of outstanding milestones for which the expense was recognized in prior years. 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 increases in prepaid insurance expenses and prepaid expenses to CROs related to our clinical trials. The decrease in operating lease liabilities is due to monthly payments for our right-of-use assets.

Investing Activities

During the nine months ended September 30, 2020 investing activities provided $92.1 million of cash, consisting of $252.3 million from proceeds of maturities of short-term investments, partially offset by $344.2 million of purchases of short-term investments and $0.2 million of purchases of property and equipment.

During the nine months ended September 30, 2019 investing activities provided $47.3 million of cash, consisting of $454.6 million from proceeds of maturities of short-term investments, partially offset by $406.1 million of purchases of short-term investments and $1.2 million of purchases of property and equipment.

Financing Activities

During the nine months ended September 30, 2020, net cash provided by financing activities was $226.4 million, consisting of net proceeds of $220.7 million from our issuance and sale of Class A common shares in a follow-on public offerings, inclusive of the exercise of the underwriters' over-allotment option to purchase additional Class A common shares, and concurrent issuance and sale of Class A1 common shares in a private placements, after deduction of

underwriting discounts and commissions, placement agent fees and other offering costs and $5.7 million of proceeds primarily from the exercise of share options.

During the nine months ended September 30, 2019, net cash provided by financing activities was $83.7 million, primarily consisting of net proceeds of $83.0 million from our issuance and sale of Class A common shares in a follow-on public offering, inclusive of the exercise in part of the underwriters' over-allotment option to purchase additional Class A common shares and concurrent issuance and sale of Class A1 common shares in a private placement, after deduction of underwriting discounts and commissions, placement agent fees and other offering costs.

Funding Requirements

We expect to incur significant expenses in connection with our ongoing and planned activities as we advance our product candidates through preclinical and clinical development, seek regulatory approval and prepare for commercial operations. 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. 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. For example, under the Regeneron Agreement, we are obligated to make payments to Regeneron of up to an aggregate of $27.5 million upon the achievement of specified regulatory milestones, including, a $7.5 million payment which may be met in the fourth quarter of 2020. Additionally, we expect to continue to incur costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses. We expect to incur expenses as we:

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

- in-license or acquire other product candidates and technologies or their related businesses, if we determine to do so.

We believe that our existing cash, cash equivalents and short-term investments will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months. We have based these estimates on assumptions that may prove to be wrong, and we could utilize our available capital resources sooner than we expect. We anticipate that we may require additional capital if we choose to pursue in-licenses or acquisitions of other product candidates and technologies or their related businesses. If we receive regulatory approval for rilonacept 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 nine months ended September 30, 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 nine months ended September 30, 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. Based on our closing share price and the market value of our voting and non-voting common shares held by non-affiliates as of September 30, 2020, we have determined that we will be a large accelerated filer and cease being an "emerging growth company" as of December 31, 2020 and cease being a

"smaller reporting company" as of January 1, 2021. After we cease being an emerging growth company and smaller reporting company, we will no longer be permitted to rely on exemptions from certain requirements that are applicable to public companies that are not emerging growth companies or smaller reporting 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 September 30, 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 September 30, 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 September 30, 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 that has 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. Our future success is dependent on our ability to develop, obtain regulatory approval for and successfully commercialize one or more of our product candidates. For example, while we announced positive data from our global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, named RHAPSODY, we have not yet demonstrated our ability to 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.

We have incurred significant losses related to expenses for research and development and our ongoing operations. As of September 30, 2020, we had an accumulated deficit of $463.8 million. We expect to continue to incur losses for the foreseeable future, and anticipate these losses will increase substantially as a result of many factors, including:

- establishing a sales, marketing and distribution infrastructure to commercialize products for which we may obtain marketing approval, including our lead program, rilonacept for approved indications in the United States;
- our research and preclinical and clinical development of our product candidates, including our global Phase 2 clinical trial with mavrilimumab for the treatment of giant cell arteritis, or GCA, for which we announced that the trial met both the primary and secondary efficacy endpoints with statistical significance, our global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe coronavirus disease 2019, or COVID-19, pneumonia and hyperinflammation, for which we are enrolling and dosing patients, and our Phase 1 clinical trial in healthy volunteers for KPL-404, as well as our planned Phase 2b dose-ranging trial with vixarelimab in prurigo nodularis;
- manufacturing our product candidates for clinical or commercial use, and increasing our manufacturing capabilities or adding additional manufacturers or suppliers;

- seeking regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;
- initiating potential additional preclinical studies and clinical trials for our product candidates;
- making milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreements;
- 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;
- 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 the impact of the COVID-19 pandemic and measures taken in response to the COVID-19 pandemic, failed trials, complex results, safety issues, regulatory challenges that require longer follow-up of existing trials, additional major trials, or additional supportive trials in order to pursue marketing approval.

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

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

We will require substantial additional financing, and a failure to obtain this necessary capital when needed on acceptable terms, or at all, will force us to delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or other operations or commercialization efforts.

The development and commercialization of biopharmaceutical products is capital intensive. We are advancing our product candidates through research, preclinical and clinical development, including our global Phase 2 clinical trial for mavrilimumab for the treatment of GCA, for which we announced that the trial met both the primary and secondary efficacy endpoints with statistical significance, the global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation, for which we are enrolling and dosing patients, and our Phase 1 clinical trial with KPL-404 in healthy volunteers, as well as our planned Phase 2b dose-ranging trial with vixarelimab in prurigo nodularis.

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. We announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY, of rilonacept in recurrent pericarditis. Upon approval from the FDA, if any, of an sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. We expect to incur significant additional commercialization expenses leading up to and after marketing approval for rilonacept or any of our other product candidates 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 profit-split or royalty payments to the licensors and other third parties from whom we have acquired our product candidates. Furthermore, we expect to continue to incur costs associated with operating as a public company, including with respect to increased compliance and disclosure obligations of becoming a large accelerated filer at the end of the year and no longer being an emerging growth company at year end or a smaller reporting company in 2021.

Accordingly, we will need to obtain substantial additional funding in connection with our continuing operations. If we are unable to raise capital when needed on acceptable terms, if at all, we will be forced to delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or commercialization efforts. We also may not be able to expand our operations or otherwise capitalize on our business opportunities, or may be required to relinquish rights to our product candidates or products.

The 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, including delays in commercialization of rilonacept or any of our other product candidates, and we may need to seek additional funds sooner than expected, through public or private securities offerings, debt financings or other sources, including government funding or grants. Our future capital requirements will depend on, and could increase significantly as a result of, many factors, including:

- our ability to successfully commercialize 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, including preparations for the potential commercial launch of rilonacept in recurrent pericarditis;

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

- the results from, and the time and cost necessary for development of our product candidates, including for our global Phase 2 clinical trial for mavrilimumab in GCA, and our global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation, and our Phase 1 clinical trial in healthy volunteers for KPL-404 as well as our planned Phase 2b dose-ranging trial of vixarelimab in prurigo nodularis;

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

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

- the costs, timing and outcomes of seeking and potentially obtaining approvals from the FDA or comparable regulatory authorities outside of the United States, including the potential for the FDA or such comparable regulatory authorities to require that we conduct more studies than we currently plan to conduct and the costs of post-marketing studies or risk evaluation and mitigation strategies, or REMS, that could be required by regulatory authorities;

- the timing and amount of milestone and other payments we must make under our agreements with Regeneron, MedImmune, Limited, or MedImmune, Biogen MA Inc., or Biogen, and the other third parties from whom we have acquired or in-licensed our product candidates or from whom we may in the future acquire or in-license product candidates;

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

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

- the receptivity of the capital markets to financings by biopharmaceutical companies generally and companies with 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 significantly 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 rilonacept is approved and marketed for human use for the treatment of CAPS in the United States by Regeneron, we are studying rilonacept for the treatment of a different indication called recurrent pericarditis, for which we announced that our global, pivotal Phase 3 clinical trial, RHAPSODY, met statistical significance on its primary and all major secondary efficacy endpoints, showing that rilonacept improved clinically meaningful outcomes associated with unmet medical need in recurrent pericarditis. 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, for which we announced that the trial met both the primary and secondary efficacy endpoints with statistical significance. Further, we are enrolling and dosing patients in the global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. We have been studying vixarelimab in prurigo nodularis, for which we released top-line data from our Phase 2a clinical trial, and are planning a Phase 2b dose-ranging trial of vixarelimab in prurigo nodularis. 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 cannot be certain that any of our product candidates will be successful in clinical trials. Further, rilonacept or any of our other product candidates may not receive regulatory approval even if we complete a successful pivotal 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:

- submission to and acceptance by the FDA of INDs and of clinical trial applications to governmental authorities outside of the United States for our product candidates to commence planned clinical trials or future clinical trials;
- successful completion of preclinical studies, including toxicology studies, biodistribution studies and minimally efficacious dose studies in animals, conducted, where applicable, under the FDA's Good Laboratory Practice, or GLP, regulations;
- successful site activation for, enrollment in, and completion of clinical trials, the design and implementation of which are agreed to by the applicable regulatory authorities, and the ability of our contract research organizations, or CROs, to successfully conduct such trials within our planned budget and timing parameters and without materially adversely impacting our trials;
- successful data from our clinical programs that support an acceptable risk-benefit profile of our product candidates for the targeted indications in the intended populations to the satisfaction of the applicable regulatory authorities;
- timely receipt, if at all, of regulatory approvals from applicable regulatory authorities and maintenance of any such approvals;
- as applicable, pediatric study plans acceptable to the FDA and comparable regulatory authorities outside of the United States, and follow through of any pediatric study commitments, including development of pediatric formulations where indicated;
- establishment and maintenance of arrangements with third-party manufacturers, as applicable, for continued clinical supply and commercial manufacturing;
- successful development of our manufacturing processes and transfer to third-party facilities to support future development activities and commercialization that are operated by contract manufacturing organizations, or CMOs, in a manner compliant with all regulatory requirements;
- successful manufacture of sufficient supplies of our product candidates within approved specifications for purity and efficacy from our facility and from our CMOs or other sole-source manufacturers in order to meet clinical or commercial demand, as applicable;
- establishment and maintenance of patent and trade secret protection or regulatory exclusivity for our product candidates;
- 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, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic, we could experience significant delays in, or an inability to, 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 Phase 2 clinical trial with mavrilimumab for the treatment of GCA for which we announced that the trial met both the primary and secondary efficacy endpoints with statistical significance, the global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation, for which we are enrolling and dosing patients, and a Phase 1 clinical trial with KPL-404 in healthy volunteers. Further, we conducted a Phase 2a clinical trial with vixarelimab in prurigo nodularis for which we announced top-line data, and plan to initiate a Phase 2b dose-ranging trial with vixarelimab in prurigo nodularis. 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, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic.

Commencing a clinical trial is subject to acceptance by the FDA of an IND or IND amendments, acceptance by European regulatory authorities of a Clinical Trial Application, or CTA, or acceptance by other applicable regulatory authorities, and finalizing the trial design based on discussions with the FDA, European regulatory authorities or other applicable regulatory authorities. We 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 or potentially in indications requiring limited doses where the theoretical risk of PAP is low. Further, based on FDA feedback we received in connection with its review and approval of an IND for our global Phase 2 clinical trial of mavrilimumab in GCA, we anticipate that to help inform the risk-benefit profile for the use of mavrilimumab in GCA, we will need to

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.

Commencing our planned clinical trials is also subject to approval by an IRB at each clinical trial site before a trial may be initiated, which approval could be delayed, rejected or suspended. Further the IRBs of the institutions in which such trials are being conducted, the Data Safety Monitoring Board for such trial or the FDA or other regulatory authorities may impose a suspension or termination of our clinical trials even after approval and initiation of trial sites due to a number of factors, including failure to conduct the clinical trial in accordance with regulatory requirements or our clinical protocols, inspection of the clinical trial operations or trial site by the FDA or other regulatory authorities, unforeseen safety issues or adverse side effects that arise in the trial, failure to demonstrate a benefit from using a drug, any of which could result in the imposition of a clinical hold, as well as changes in governmental regulations or administrative actions or lack of adequate funding to continue the clinical trial.

Successful completion of our clinical trials is a prerequisite to submitting a Biologics License Application, or BLA, sBLA or New Drug Applications, or NDA, to the FDA and a marketing authorization application, or MAA, to the European Medicines Agency, or EMA, or other applicable regulatory authorities in other countries for each product candidate and, consequently, to obtaining approval and initiating commercial marketing of our current and any future product candidates. 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 have and may continue to experience delays in our ongoing clinical trials, and we do not know whether planned clinical trials will begin on time, be allowed by regulatory authorities, need to be redesigned, or if we can activate sites or enroll patients on time, or if they will be completed on schedule, if at all. Events that 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 as a result of the COVID-19 pandemic and measures taken in response to the pandemic or otherwise, among other things;
- clinical trial sites or patients withdrawing from a clinical trial, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic or otherwise, among other things;
- participating patients experiencing serious adverse events or undesirable side effects or being exposed to unacceptable health risks;
- participating patients failing to experience confirmed pre-specified events during the clinical trial within an expected time-frame, if at all;
- safety issues, including occurrence of adverse events associated with the product candidate that are viewed to outweigh its potential benefits;
- difficulty in identifying the patient populations that we are trying to enroll in a particular trial, which may delay enrollment and reduce the power of a clinical trial to detect statistically significant results;
- changes in regulatory requirements, policies and guidance that require amending or submitting new clinical protocols;
- the cost of clinical trials of our product candidates being greater than we anticipate;
- clinical trials of our product candidates producing negative, inconclusive or uncompetitive results, which may result in us deciding, or regulators requiring us, to conduct additional clinical trials or modify or cease development programs for our product candidates;
- suspensions or terminations of our clinical trials by us or the IRBs of the institutions in which our clinical trials are being conducted, the Data Safety Monitoring Board for such trials or the FDA or comparable regulatory authorities;
- failure of manufacturers, or us, to produce phase-appropriate supplies of our product candidates for use in our clinical trials in accordance with current good manufacturing practices, or cGMP, requirements and regulations or applicable comparable regulatory guidelines in other countries;
- delays in manufacturing, testing, releasing, validating or importing/exporting sufficient stable quantities of our product candidates for use in clinical trials or the inability to do any of the foregoing; and
- disruptions to our business operations, including our manufacturing operations, and the business operations of our third-party manufacturers, CROs upon whom we rely to conduct our clinical trials, or other third parties with whom we conduct business or otherwise engage, as well as disruptions in travel into and within the countries in which we conduct our clinical trials, our manufacturers produce our product candidates or we otherwise conduct business or engage with other third parties, now or in the future as a result of the impact of the COVID-19 pandemic.

In addition, disruptions caused by the COVID-19 pandemic may increase the likelihood that we encounter such difficulties or delays in commencing or completing our planned and ongoing clinical trials and other clinical development. 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 significant 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 have an adverse impact on our current or planned preclinical studies and clinical trials, which could be significant.

The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact our current or planned preclinical studies and clinical trials, which could be significant. If the COVID-19 pandemic and measures undertaken in response to the pandemic are prolonged, or the easing of any of such measures has adverse consequences, we may experience significant disruptions that could 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 the supply of raw materials or global shipping that may affect the transport of our product candidates or clinical trial materials, or the reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19;

- impeding, delaying, limiting or preventing the production, delivery or release of the 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;

- 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;

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

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

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

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

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

Any one of the foregoing could significantly impede, delay, limit or prevent the clinical development of our product candidates and ultimately lead to the delay or denial of regulatory approval of our product candidates. For example, the large number of competitive studies for the treatment of conditions associated with COVID-19 and the evolving standard of care with overall improvement in the quality of care for patients diagnosed with COVID-19 are having an impact on the number of eligible patients for our the global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. While we continuously look to identify business-critical activities and to develop contingencies and mitigation strategies for those activities to potentially minimize the impact of the COVID-19 pandemic on our business and operations, there can be no assurance that we will be able to identify all such activities or that any identified contingencies and mitigation strategies

will be effective. If the clinical development of our product candidates is significantly impeded, delayed, limited or is prevented, it could ultimately lead to the delay or denial of regulatory approval of our product candidates which would materially adversely affect our business and operations, including our ability to generate revenue.

We may find it difficult to enroll patients in our clinical trials in a timely manner given the limited number of patients who have the diseases for which our product candidates are being studied, our particular enrollment criteria or competing clinical studies in the same patient population, including patients with COVID-19, or due to the impact of the COVID-19 pandemic. Difficulty in enrolling patients could delay or prevent completion of our clinical trials of our product candidates, and our research and development efforts could be adversely affected.

Identifying and qualifying patients to participate in clinical trials of our product candidates is critical to our success. The timing of our clinical trials depends in part on the speed at which we can recruit a sufficient number of patients to participate in testing our product candidates, including for the treatment of conditions associated with COVID-19, particularly given that many of the conditions for which we are evaluating our current product candidates or may evaluate them in the future are in small disease populations. In addition, the eligibility criteria of our clinical trials will further limit the pool of available trial participants, as we will require patients to have specific characteristics that we can evaluate based on the primary and secondary endpoints of the study. Further, our product candidates modulate the immune system and carry risks associated with immunosuppression, including the risk of serious infections and other potential serious health risks.

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. For example, the large number of competitive studies for the treatment of conditions associated with COVID-19 and the evolving standard of care with overall improvement in the quality of care for patients diagnosed with COVID-19 are having an impact on the number of eligible patients for our global placebo-controlled Phase 2 portion of our adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. Since the number of qualified clinical investigators is limited, we may conduct some of our clinical trials at the same clinical trial sites that some of our competitors use, which would reduce the number of patients who are available for our clinical trials at such clinical trial site.

In addition, disruptions to our business operations and the business operations of our CROs or other third parties with whom we conduct business, as well as disruptions in travel into and within the countries in which we conduct our clinical trials, now or in the future, as a result of the impact of the COVID-19 pandemic, may delay or prevent patient enrollment. For example, our CRO for the Phase 1 clinical trial of KPL-404 notified us that, after a temporary pause due to the impact of the COVID-19 pandemic, they resumed certain clinical trial activities, but we also engaged an additional CRO to conduct additional portions of the trial at another clinical trial site.

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 and evolving standards of care;
- the proximity of patients to clinical sites;
- the complexity of the design and nature of the clinical protocol and trial;

- the availability and nature of competing clinical trials;
- the availability of standard of care or new drugs approved for the indication the clinical trial is investigating;
- failure to obtain and maintain or timely amend patient consents;
- our ability to recruit clinical trial investigators with applicable competencies and experience;
- the risk that patients enrolled in clinical trials will withdraw from the trials before completion of their treatment or follow-up period (in either case including as a result of trial demands on participants among other things);
- clinicians' and patients' perceptions as to the safety and potential advantages of the product candidate being studied in relation to other available therapies; and
- the occurrence of adverse events, or AEs, or undesirable side effects attributable to our product candidates.

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

Our 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 rilonacept 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 rilonacept. In our open-label Phase 2 proof-of-concept clinical trial of rilonacept for recurrent pericarditis, the most common AEs were gastrointestinal disorders and injection site reactions and there was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment. In our global, pivotal Phase 3 clinical trial, RHAPSODY, the most common AEs were injection site reactions and upper respiratory tract infections and there were four serious AEs on rilonacept, none of which were treatment-related.

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 or potentially in indications requiring limited doses where the theoretical risk of PAP is low. In addition, based on FDA feedback we received in connection with its review and authorization of an IND for our global Phase 2 clinical trial of mavrilimumab in GCA, we anticipate that to help inform the risk-benefit profile for the use of mavrilimumab in GCA, we will need to 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 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.

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

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 third parties, including the companies from whom we have licensed or acquired or may in the future license or acquire our product candidates, in our later clinical trials, we may be unable to successfully develop, obtain regulatory approval for and commercialize our product candidates.

Positive results from our preclinical studies and any positive results we may obtain from our earlier clinical trials of our product candidates, or from the clinical trials conducted by third parties, including investigator-initiated studies or the companies from whom we in-licensed or acquired or may in the future in-license or acquire our product candidates, may not be predictive of the results from any required later preclinical studies and clinical trials. Similarly, the positive results from the preclinical studies and earlier clinical trials or investigator initiated studies of our product candidates may not be replicated in our subsequent preclinical studies and clinical trial or investigator initiated study results. The mechanisms of action of our product candidates may not prove to be safe or effective to treat the diseases we are studying. Further, the safety and efficacy of our product candidates have not been determined for the indications in which we are developing them, and we cannot provide any assurance that their development will be successful. For example, although rilonacept is FDA approved for the treatment of CAPS, and we announced that our global, pivotal Phase 3 clinical trial, RHAPSODY, with rilonacept in recurrent pericarditis met statistical significance on its primary and all major secondary efficacy endpoints, and mavrilimumab has been studied in Phase 2 clinical trials for the treatment of RA by MedImmune, and we announced that our global Phase 2 clinical trial for the treatment of GCA met achieved statistical significance on both its primary and secondary efficacy endpoints, their safety and efficacy have not been determined in the indications we are pursuing, 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 preclinical studies and clinical trials. Moreover, preclinical and clinical data are often susceptible to varying interpretations and analyses and many companies that believed their product candidates performed satisfactorily in preclinical studies and clinical trials nonetheless failed to obtain FDA or EMA approval. Furthermore, the approval policies or regulations of the FDA or the applicable regulatory authorities outside of the United States may significantly change in a manner rendering our clinical data insufficient for approval, which may lead to the FDA or such other regulatory authorities delaying, limiting or denying approval of our product candidates.

Interim, preliminary, and "top-line" data from our clinical trials that we announce or publish from time to time may change as more patient data become available following the interim data; preliminary data are subject to audit and

verification procedures, and deeper analysis of the data beyond the topline data may provide more color and context to the data, all of which could result in material or other changes in the final data.

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

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

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

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

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

Risks Related to Marketing Approval and Regulatory Matters

Regulatory approval processes are lengthy, time consuming and inherently unpredictable. If we are not able to obtain, or if there are delays in obtaining, required regulatory approvals for our 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, record-keeping, labeling, storage, approval, advertising, promotion, pricing, sale, distribution, import and export are subject to comprehensive regulation by the FDA and other regulatory agencies in the United States and by comparable authorities in other countries. Before we can commercialize any of our 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 authorities. 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, including with respect to approval for the commercial marketing of rilonacept in the United States for recurrent pericarditis.

We announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY, of rilonacept in recurrent pericarditis. Upon approval from the FDA, if any, of an sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States. We must continue to coordinate numerous activities with Regeneron in order for us to take over certain responsibilities and obligations with respect to owning Regeneron's BLA for CAPS. The activities related to ensuring our readiness to take over such responsibilities and obligations with respect to owning the BLA are complex and resource intensive, and we may not have anticipated all such activities to be coordinated or responsibilities and obligations to be assumed. Further, if Regeneron or we do not perform in accordance with our expectations, or we are not ready to assume the responsibilities and obligations on a timely basis, or either we or Regeneron have critical or major observations during the FDA's anticipated pre-approval inspection, or PAI, with respect to our and Regeneron's ability to manufacture rilonacept to the requisite level of quality standards and controls could delay FDA approval or our commercialization of rilonacept, and our ability to generate revenue from rilonacept could be materially impaired. In addition, we could have critical or major observations during the FDA's anticipated

Bioresearch Monitoring program, or BIMO, inspection as to the quality and integrity of the clinical trial conduct and of data submitted to the FDA under an sBLA for rilonacept, or if either inspection is delayed, any of which could delay FDA approval or our commercialization of rilonacept, and our ability to generate revenue from rilonacept 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;
- the FDA or comparable regulatory authorities in other jurisdictions could require us to collect additional data or conduct additional clinical trials; 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;
- 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, sBLA or other submission or to obtain regulatory approval in the United States or elsewhere;
- the FDA or comparable regulatory authorities in other jurisdictions could require us to conduct additional clinical trials to compare our product candidates to other therapies for the treatment of the same indication;
- the number of patients required for clinical trials of our product candidates may be larger than we anticipate, enrollment in these clinical trials may be slower than we anticipate, participants may drop out of these clinical trials at a higher rate than we anticipate or we may fail to recruit suitable patients to participate in a trial;

- our third-party contractors may fail to comply with regulatory requirements or meet their contractual obligations to us in a timely manner, or at all;

- the FDA or comparable regulatory authorities in other jurisdictions may not believe that we have sufficiently demonstrated our ability to manufacture the products to the requisite level of quality standards, or they may fail to approve our manufacturing processes or facilities, or the manufacturing processes or facilities of third-party manufacturers with which we contract for clinical and commercial supplies;

- the FDA or comparable regulatory authorities in other jurisdictions may not believe that their on-site inspections and data audits have sufficiently demonstrated the quality and integrity of the clinical trial conduct and of data submitted to the FDA or comparable regulatory authorities in other jurisdictions in support of our new product approvals and marketing applications;

- the supply or quality of our product candidates or other materials necessary to conduct clinical trials of our product candidates may be insufficient or inadequate;

- our product candidates may have undesirable side effects, toxicities or other unexpected characteristics, causing us or our investigators, regulators or IRBs to reject, suspend or terminate the clinical trials; and

- the approval policies or regulations of the FDA or comparable regulatory authorities in other jurisdictions may significantly change in a manner rendering our clinical data, biologic manufacturing process and other supporting information insufficient for approval.

In addition, even if we were to obtain approval for one or more of our 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, although rilonacept was approved as a biological product under a BLA for the treatment of CAPS in February 2008, and we believe it qualified for the 12-year period of exclusivity against any biosimilars, such 12-year period of exclusivity has lapsed. We announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY, of rilonacept in recurrent pericarditis. Upon approval from the FDA, if any, of a sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States. However, the 12-year exclusivity period does not attach to the approval of an sBLA, potentially creating the opportunity for biosimilar competition, subject to any orphan drug exclusivity under the U.S. Orphan Drug Act (see "Risks Related to Marketing Approval and Regulatory Matters - We received orphan drug designation in the United States for rilonacept for the treatment of pericarditis and for mavrilimumab for the treatment of GCA and may seek orphan drug designation for some of our other product candidates in the United States, as well as for any of our product candidates in the EU, and we may be unsuccessful, or may be unable to maintain the benefits associated with orphan drug designation, including the potential for market exclusivity, for any product candidate for which we obtain orphan drug designation"). If we obtain FDA approval for any of our other biological product candidates, we expect any such product candidates to qualify for the 12-year period of exclusivity under the BPCIA. However, there is a risk that this exclusivity could be shortened due to Congressional action or otherwise, or that the FDA will not consider any such approved product candidates, to be reference products for competing products, potentially creating the opportunity for generic competition sooner than anticipated.

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

Even if we obtain marketing 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 received orphan drug designation in the United States for rilonacept for the treatment of pericarditis and for mavrilimumab for the treatment of GCA and may seek orphan drug designation for some of our other product candidates in the United States, as well as for any of our product candidates in the EU, and we may be unsuccessful, or may be unable to maintain the benefits associated with orphan drug designation, including the potential for market exclusivity, for any product candidate for which we obtain orphan drug designation.

We received orphan drug designations in the United States for rilonacept for the treatment of pericarditis, which includes recurrent pericarditis and for mavrilimumab for the treatment of GCA, and we may seek orphan drug designation for certain of our other product candidates in the United States as well as for any of our product candidates in the EU. We may be unsuccessful in obtaining such designation for any of our other product candidates or unable to maintain the associated benefits for rilonacept or mavrilimumab or any of our other product candidates that are granted orphan drug designation, if any. Regulatory authorities in some jurisdictions, including the United States and Europe, may designate drugs or biologics intended to treat relatively small patient populations as orphan drug products. Under the U.S. Orphan Drug Act, the FDA may designate a drug or biologic as an orphan drug if it is intended to treat a rare disease or condition, which is generally defined as a patient population of fewer than 200,000 individuals in the United

States, or a patient population greater than 200,000 in the United States where there is no reasonable expectation that the cost of developing the drug will be recovered from sales in the United States.

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

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

Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent the FDA from approving the same or a different drug in another indication. Even after an orphan drug is approved, the FDA can subsequently approve a later application for the same drug for the same condition if the FDA concludes that the later drug is clinically superior in that it is shown to be safer in a substantial portion of the target populations, more effective or makes a major contribution to patient care. In addition, a designated orphan drug may not receive orphan drug exclusivity if it is approved for a use that is broader than the indication for which it received orphan designation. Moreover, orphan-drug-exclusive marketing rights in the United States may be lost if the FDA later determines that the request for designation was materially defective or if we are unable to manufacture sufficient quantities of the product to meet the needs of patients with the rare disease or condition. Orphan drug designation neither shortens the development time or regulatory review time of a drug nor gives the drug any advantage in the regulatory review or approval process. While we received orphan drug designations in the United States for rilonacept for the treatment of pericarditis and for mavrilimumab for the treatment of GCA, and we may seek orphan drug designation for certain of our product candidates, we may never receive such designation for such other product candidates. Even though we received such designation for rilonacept and mavrilimumab and may receive such designation for any of our other product candidates, there is no guarantee that we will enjoy the benefits of such designations.

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

We received Breakthrough Therapy designation for rilonacept for the treatment of recurrent pericarditis, and we may seek Breakthrough Therapy or Fast Track designation for some of our other product candidates. A Breakthrough Therapy is defined as a drug that is intended, alone or in combination with one or more other drugs or biologics, to treat a serious or life-threatening disease or condition, and preliminary clinical evidence indicates that the drug may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. In addition, if a product candidate is intended for the

treatment of a serious or life-threatening condition and clinical or preclinical data demonstrate the potential to address unmet medical needs for this condition, the sponsor may apply for Fast Track designation.

The FDA has broad discretion whether or not to grant Fast Track and Breakthrough Therapy designations, and even if we believe a particular product candidate is eligible for this designation, we cannot be certain that the FDA would decide to grant it. Even if we obtain such designations for one or more of our product candidates, we may not experience a faster development process, review or approval compared to non-expedited FDA review procedures. In addition, the FDA may withdraw Fast Track or Breakthrough Therapy designations if it believes that such designations are no longer supported. Although products receiving Fast Track and Breakthrough Therapy designation are generally eligible for the FDA's priority review procedures, receiving such designations does not guarantee that the BLA for such products will receive priority review.

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

We have never 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 a pivotal clinical trial or obtain marketing approval 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 our global, pivotal Phase 3 clinical trial, RHAPSODY, for rilonacept in recurrent pericarditis and members of our management team have obtained marketing approvals for product candidates in the past while employed at other companies, we as a company have not obtained marketing approvals for any of our product candidates. As a result, such activities may require more time than we anticipate. Failure to successfully complete, or delays in, any of our eventual other pivotal trials or related regulatory submissions would prevent us from, or delay us in, obtaining regulatory approval for, or clearance of, our product candidates. It is possible that the FDA or other government agencies may refuse to accept for substantive review any regulatory submissions that we submit for rilonacept or any of our other product candidates or may conclude after review of our applications for rilonacept or any of our other product candidates that the submissions are insufficient to obtain marketing approval or clearance of rilonacept or any of our other product candidates. For example, we announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY, of rilonacept in recurrent pericarditis. If the FDA or other government agencies do not accept our applications or issue marketing authorizations for rilonacept or any of our other 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 rilonacept or any of our other 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 one or more of our product candidates, which could significantly harm our business.

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

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

Separately, in response to the COVID-19 pandemic, on March 10, 2020 the FDA announced its intention to postpone most inspections of foreign manufacturing facilities, and on March 18, 2020, the FDA temporarily postponed routine surveillance inspections of domestic manufacturing facilities. Subsequently, on July 10, 2020 the FDA announced its intention to resume certain on-site inspections of domestic manufacturing facilities subject to a risk-based prioritization system. The FDA intends to use this risk-based assessment system to identify the categories of regulatory activity that can occur within a given geographic area, ranging from mission-critical inspections to resumption of all regulatory activities. Regulatory authorities outside the United States may adopt similar restrictions or other policy measures in response to the COVID-19 pandemic. If a prolonged government shutdown occurs, or if global health concerns continue to prevent the FDA or other regulatory authorities from conducting their regular inspections, reviews, or other regulatory activities, it could significantly impact the ability of the FDA or other regulatory authorities to timely review and process our regulatory submissions, which could have a material adverse effect on our business.

Risks Related to Manufacturing and Our Reliance on Third Parties

We *contract with third parties for manufacturing our 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, preclinical and other clinical development for our product candidates, we rely, and expect to continue to rely, on third parties for the manufacture of our late-stage product candidates and certain of our early-stage product candidates for the majority of our clinical development efforts, as well as for the potential commercial manufacture of our product candidates, if approved, as well as label and packaging activities. We rely on these third parties 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, Regeneron is the sole manufacturer of rilonacept and we have a contract with Regeneron to produce rilonacept on an exclusive basis for a period of time. Regeneron, in turn, relies upon a third party CMO to conduct fill/finish operations for rilonacept. Under certain circumstances, we or Regeneron could initiate a technology transfer to either us or another CMO to manufacture rilonacept. Finding new CMOs or third-party suppliers to produce rilonacept would add additional cost and require significant time and focus of our management team. The CMO would need to produce rilonacept at a different manufacturing site and potentially using a different process or at a different scale. We cannot provide any assurance that the technology transfer from Regeneron to us or another CMO will be successful in producing rilonacept in sufficient quantities or of acceptable quality, if at all, or that it will produce a comparable product to the satisfaction of the FDA or other comparable regulatory authorities, which could delay, prevent or impair the development or commercialization of rilonacept. In addition, there is typically a transition period when a new CMO commences work. Any significant delay or interruption in the supply of rilonacept could considerably impact our ability to meet the clinical trial or commercial demand for the product and our ability to generate revenue from rilonacept could be materially impaired.

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

As a result of the COVID-19 pandemic, existing and any new third party CMOs or suppliers may be unable to produce or supply our product candidates or the raw materials needed to produce or supply our product candidates or may experience delays, restrictions or limitations in the production, delivery or release of the supply of our product candidates or the raw materials needed to produce our product candidates, including due to disruptions at the respective facilities that produce our product candidates or the raw materials needed to produce our product candidates, staffing shortages or reprioritizations, production slowdowns, stoppages or reprioritizations, including as a result of reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, or interruptions in global shipping. In addition, there is typically a transition period when a new CMO commences work. Finding new CMOs or third-party suppliers involves additional cost and requires our management's time and focus. 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.

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. In addition, as we plan to produce clinical trial and potential commercial material at a CMO, the CMO may be required to adopt different manufacturing protocols or processes. For example, although Regeneron has produced rilonacept for commercial use for over ten years, the FDA or other applicable regulatory authorities in other jurisdictions may reevaluate rilonacept's current manufacturing processes or route of administration in connection with evaluating whether to approve rilonacept for a new indication, such as recurrent pericarditis.

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.

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, including as a result of the COVID-19 pandemic, could cause product shortages, disrupt or delay our clinical trials or regulatory approvals, delay or stop commercialization of our 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, and 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, shipping 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.

Any adverse developments affecting the operations of our third-party providers, such as any impact due to the COVID-19 pandemic including shortages or reprioritizations of raw materials, including reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, could result in a shortage of clinical products or impose commercial product requirements, cause 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 rilonacept, mavrilimumab and vixarelimab are supplied to us from single-source suppliers. Regeneron has a contractual right to be our sole source manufacturer of rilonacept 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 rilonacept, mavrilimumab and vixarelimab in the quantities that we believe would be required to meet anticipated market demand, our third-party manufacturers may need to increase manufacturing capacity and, in some cases, alternative sources of commercial supply may need to be secured, which could involve significant challenges and may require additional regulatory approvals. In addition, the development of commercial-scale manufacturing capabilities may require us and our third-party manufacturers to invest substantial additional funds and hire and retain the technical personnel who have the necessary manufacturing experience. Neither we nor our third-party manufacturers may successfully complete any required increase to existing manufacturing capacity in a timely manner, or at all.

Moreover, if there is a disruption to our manufacturing operations or one or more of our third-party manufacturers' or suppliers' relevant operations, such as due to the impact of the COVID-19 pandemic, including due to staffing shortages or reprioritizations, production slowdowns or stoppages or interruptions in global shipping, the supply of the related product candidate will be delayed until we or such manufacturer or supplier restores the affected facilities or we or they procure alternative manufacturing facilities or sources of supply. Our ability to progress our preclinical and clinical programs 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 activate sites, conduct and otherwise support our research activities, preclinical studies, clinical trials and other trials for our product candidates. If these third parties do not successfully carry out their contractual duties, comply with regulatory requirements or meet expected deadlines, we may not be able to obtain regulatory approval for or commercialize our product candidates, and our business could be substantially harmed.

We rely on medical institutions, clinical investigators, contract laboratories and other third parties, such as CROs, to activate sites, conduct or otherwise support our 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 such site activation, execution of and otherwise supporting 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 preclinical studies or clinical trials, we could be subject to warning letters or enforcement actions that may include civil penalties and criminal prosecution.

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

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

- have staffing difficulties;
- have disruptions to their business and operations, including as a result of the impact from an epidemic or pandemic disease outbreak, including COVID-19 (see "Risk Factors — Risks related to product development — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an

adverse impact on our current or planned preclinical studies and clinical trials, which could be significant.");

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

These factors may materially adversely affect the willingness or ability of third parties to activate sites and conduct and oversee our clinical trials and may subject us to unexpected cost increases that are beyond our control. If the CROs, their subcontractors or the clinical trial sites do not perform clinical trials in a satisfactory manner, breach their obligations to us or fail to comply with regulatory requirements, the development, regulatory approval and commercialization of our product candidates may be delayed, we may not be able to obtain regulatory approval and commercialize our product candidates, or our development program may be materially and irreversibly harmed. If we are unable to rely on clinical data collected by our CROs, their subcontractors or the clinical trial sites, we could be required to repeat, extend the duration of or increase the size of any clinical trials we conduct and this could significantly delay commercialization and require significantly greater expenditures.

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

If the third parties conducting our preclinical studies or our clinical trials do not perform their contractual duties or obligations, experience significant business challenges, disruptions or failures, such as due to the impact of the COVID-19 pandemic, do not meet expected deadlines, terminate their agreements with us or need to be replaced, or if the quality or accuracy of the data they obtain is compromised due to their failure to adhere to our protocols or to GCPs, or for any other reason, we may need to enter into new arrangements with alternative third parties. This could be difficult, costly or impossible, and our preclinical studies or clinical trials may need to be extended, delayed, terminated or repeated. As a result, we may not be able to obtain regulatory approval in a timely fashion, or at all, for the applicable product candidate, our financial results and the commercial prospects for our product candidates would be harmed, our costs could increase, and our ability to generate revenues could be delayed.

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

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

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

Risks Related to Competition, Executing our Strategy, Retaining Key Employees and Managing Growth

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

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

While we are not aware of any therapies currently approved or actively continuing clinical trials in recurrent pericarditis, there is one product that modulates the signaling of IL-1α and IL-1β, anakinra (KINERET), marketed by Swedish Orphan Biovitrum AB, and one product that modulates the signaling of IL-1β, canakinumab (ILARIS), marketed by Novartis Pharmaceuticals Corporation. There are other therapies which modulates IL-1α in preclinical and clinical development for diseases other than recurrent pericarditis from Johnson & Johnson and XBIOTECH USA, INC. We expect mavrilimumab, if approved for the treatment of GCA, to experience competitive pressure from tocilizumab (ACTEMRA), marketed by Genentech USA, Inc., which was approved in 2017 for use in GCA as an adjunct to steroid taper. Additional competition may be experienced from Eli Lilly and Company and AbbVie Inc., which are conducting clinical trials for oral janus kinase inhibitors, Sanofi S.A. and Regeneron, which are recruiting a Phase 3 clinical trial with their anti-IL-6 program, Novartis International AG, which is recruiting a trial with its IL-17 antagonist secukinumab (Cosentyx) and Janssen Biotech, Inc., which is testing ustekinumab (STELARA) in two small studies for GCA. There are multiple other programs targeting GM-CSF antagonism not currently pursuing GCA in clinical trials that could decide in the future to engage in development of therapies for GCA, including GlaxoSmithKline plc, Izana Bioscience, Roivant Sciences Ltd., I-Mab Biopharma Co., Ltd., and Humanigen, Inc. We are also pursuing mavrilimumab development for the treatment of COVID-19 pneumonia and hyperinflammation. There are currently hundreds of active, industry sponsored clinical trials testing many different mechanisms of action for the treatment of COVID-19 related therapeutic areas in addition to the many clinical trials testing vaccines for the prevention of COVID-19. Multiple therapies are in development for prurigo nodularis, and any that receive FDA approval for this indication will be likely competitors to vixarelimab. These products include nemolizumab, dupilumab and nalbuphine ER. There are multiple agents targeting antagonism of the CD40/CD40L interaction across a variety of clinical uses including, Novartis International AG, Biogen Inc., or Biogen, and UCB, Inc., C.H. Boehringer Sohn AG & Ko. KG and AbbVie Inc., 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.

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

- may have significant discretion in determining the efforts and resources that they will apply;
- may not commit sufficient resources to or otherwise not perform satisfactorily in carrying out its activities;
- may not properly maintain or defend our intellectual property rights or may use our intellectual property or proprietary information in a way that gives rise to actual or threatened litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential liability;
- may own or co-own intellectual property covering products that results from our arrangement with them, and in such cases, we would not have the exclusive right to develop or commercialize such intellectual property, and even if we are able to license such exclusive rights, we may have to enter into a license agreement that include obligations to make milestone, royalty or other payments under such agreement; and
- may conduct sales and marketing activities or other operations that may not be in compliance with applicable laws, resulting in civil or criminal proceedings.

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

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

We 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. Our Chief Financial Officer, departed the Company in March 2020, and we may not seek to find a replacement or may experience difficulties or delays in identifying a qualified replacement if we do seek to do so. 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 that expires 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 obtained orphan drug designations from the FDA for rilonacept for the treatment of pericarditis, which includes the treatment of recurrent pericarditis, and for mavrilimumab for the treatment of GCA, we may pursue orphan drug designation for our other product candidates in the United States and we may not be successful in obtaining such designation, or we may not be able to maintain the benefits of the designation for rilonacept or mavrilimumab or any of our other product candidates. Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent the FDA from approving the same or a different drug in another indication. Even after an orphan drug is approved, the FDA can subsequently approve a later application for the same drug for the same condition if the FDA concludes that the later drug is clinically superior in that it is shown to be safer in a substantial portion of the target populations, more effective or makes a major contribution to patient care. In addition, a designated orphan drug may not receive orphan drug exclusivity if it is approved for a use that is broader than the indication for which it received orphan designation. See "Risk Factors — Risks related to marketing approval and regulatory matters."

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

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

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

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

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

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

be upheld in the face of a potential challenge, or that they will not be breached, for which we may not have an adequate remedy.

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

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

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

We entered into agreements to acquire the rights to develop or commercialize our product candidates, rilonacept, 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 rilonacept. 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. We announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY, of rilonacept in recurrent pericarditis. Upon receipt of FDA approval for rilonacept in recurrent pericarditis, if any, we would assume the sales and distribution of rilonacept for the other approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. We must continue to coordinate numerous activities with Regeneron in order for us to take over certain responsibilities and obligations with respect to owning the BLA. 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, or DIRA. 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 Biogen Agreement.

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

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

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

Since our product candidates are being developed for use in fields that are competitive and of strong interest to pharmaceutical and biotechnology companies, we will likely seek to file additional patent applications and may have

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

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

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

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

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

An adverse result in any litigation proceeding could put one or more of our patents, whether owned or in-licensed, at risk of being invalidated or interpreted narrowly. If a defendant were to prevail on a legal assertion of invalidity or unenforceability of our patents covering one of our product candidates, we would lose at least part, and

perhaps all, of the patent protection covering such product candidate. Competing products may also be sold in other countries in which our patent coverage might not exist or be as strong. If we lose a patent lawsuit outside of the United States, alleging our infringement of a competitor's patents, we could be prevented from marketing our products in one or more such countries. Any of these outcomes would have a materially adverse effect on our business.

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

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

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

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

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

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

Filing, prosecuting and defending patents on our product candidates in all countries throughout the world would be prohibitively expensive. The requirements for patentability may differ in certain countries, particularly in developing countries. Moreover, our ability to protect and enforce our intellectual property rights may be adversely affected by unforeseen changes in intellectual property laws outside of the United States. In addition, the patent laws of some such countries do not afford intellectual property protection to the same extent as the laws of the United States. Many companies have encountered significant problems in protecting and defending intellectual property rights in certain jurisdictions outside of the United States. Varying filing dates in international countries may also permit intervening

third parties to allege priority to patent applications claiming certain technology. The legal systems of some countries, particularly developing countries, do not favor the enforcement of patents and other intellectual property rights. This could make it difficult for us to stop the infringement of our patents or the misappropriation of our other intellectual property rights. For example, many countries outside of the United States have compulsory licensing laws under which a patent owner must grant licenses to third parties. In addition, many countries limit the enforceability of patents against certain parties, including government agencies or government contractors. Consequently, we may not be able to prevent third parties from practicing our inventions in all countries outside the United States. Competitors may use our technologies in jurisdictions where we have not obtained patent protection to develop their own products and, further, may export otherwise infringing products to territories where we have patent protection, if our ability to enforce our patents to stop infringing activities is inadequate. These products may compete with our product candidates, and our patents or other intellectual property rights may not be effective or sufficient to prevent them from competing.

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

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

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

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

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

patents and patents that we might obtain in the future; shorten the term that has been lengthened by patent term adjustment of our existing patents or patents that we might obtain in the future; or challenge the validity or enforceability of our patents that may be asserted against us by our competitors or other third parties. Any of these outcomes could have a material adverse effect on our business.

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

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

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

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

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

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

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

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

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

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, including with respect to COVID-19 pneumonia and hyperinflammation. Our projections of both the number of people who have these diseases, as well as the subset of people with these diseases who have the potential to benefit from treatment with our product candidates, are based on our extrapolation from available population data and estimates. These estimates have been derived from a variety of sources, including scientific literature, surveys of clinics, patient foundations, pharmacy claims analyses, large national surveillance databases or market research, and may prove to be incorrect. Further, new trials and therapeutic options may lead to changes in the estimated incidence or prevalence of these diseases, or relevant subpopulations thereof, including the introduction of a vaccine for the prevention of COVID-19 with respect to COVID-19 pneumonia and hyperinflammation. As a result, the number of patients who may benefit from our 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 as a company. 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, distribution, access and physician and patient support capabilities as well as the infrastructure to support our operations now and into the future in order to directly commercialize rilonacept in the United States, 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 develop strong scientific-based relationships to drive disease awareness and education;
- 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 provide physicians and patients adequate support and training to build comfort around reconstitution and self-administration process to initiate our products;
- 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.

In addition, disruptions caused by the COVID-19 pandemic may increase the likelihood that we encounter such difficulties, delays or unforeseen costs. 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 if not completed as planned could delay the launch of our product candidates. We may not be able to adequately build an effective sales, marketing and access organization in the United States, the EU or other key markets in which we have obtained approval for the commercial marketing of our product candidates. 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, and conducting in-person market research as well as recruiting qualified candidates to enhance our commercial operations and support commercialization, which, if prolonged, may impede the effective commercialization of our product candidates and result in lower than anticipated future revenue.

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 rilonacept or any of our other 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 rilonacept or any of our other product candidates, or effectively block or limit their use in the case of third-party payors. For example, we announced that our global, pivotal Phase 3 clinical trial, RHAPSODY of rilonacept for the treatment of recurrent pericarditis, met statistical significance on its primary and all major secondary efficacy endpoints, showing that rilonacept improved clinically meaningful outcomes associated with unmet medical need in recurrent pericarditis. Upon approval from the FDA, if any, of an sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. If rilonacept in recurrent pericarditis or any of our other 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 rilonacept, if approved, or any of our other product candidates will depend on a variety of factors, including:

- the timing of market introduction;
- disease awareness;
- 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 the timing thereof; and
- other potential advantages over alternative treatment methods.

If rilonacept in recurrent pericarditis or any of our other product candidates fail to gain market acceptance, our ability to generate revenue will be adversely affected. Even if rilonacept or any of our other product candidates achieve market acceptance, the relevant market may prove not to be large enough to allow us to generate significant revenue.

The successful commercialization of our 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 rilonacept in recurrent pericarditis or any of our other 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). Although rilonacept is approved and marketed for human use for the treatment of CAPS in the United States by Regeneron, if rilonacept is approved by the FDA for the commercial marketing of rilonacept in the United States for recurrent pericarditis we would need to seek favorable coverage and reimbursement for this indication from third party payors. 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 and therapeutics. To commercialize rilonacept in recurrent pericarditis, if approved, or any of our other product candidates successfully, we will be required to have sufficient expertise, internally or through a third party, and sufficient resources to execute on the respective product candidate’s coverage and reimbursement strategy. We cannot be certain we will be able to timely and effectively execute our coverage and

reimbursement strategy in the markets we pursue, which could limit the commercial potential of rilonacept in recurrent pericarditis or any of our other product candidates and our ability to generate projected revenue from rilonacept or any of our other product candidates.

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 methodologies, 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 methodologies 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 of the countries we may pursue outside of the United States for any of our product candidates, the products may be subject to extensive governmental price control or other price regulations. Some countries require approval of the sale price of a drug before it can be marketed. In many countries, the pricing review period begins after marketing approval is granted. In some markets, prescription pharmaceutical pricing remains subject to continuing governmental control even after initial approval is granted. As a result, we might obtain marketing approval for a product candidate in a particular country, but then be subject to price negotiations that delay our commercial launch of the product candidate in that country, possibly for lengthy time periods, which may negatively impact the revenues we are able to generate from the sale of the product candidate in that country. Adverse pricing and

reimbursement limitations may hinder our ability to recoup our investment in one or more product candidates, even if our product candidates obtain marketing approval.

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

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

Although we do not have immediate plans to pursue the commercialization of rilonacept for recurrent pericarditis or any other approved indication outside of the United States, we are evaluating the opportunities for the development and commercialization of our product candidates in certain markets outside of the United States. We are not permitted to market or promote any of our product candidates before we receive regulatory approval from the applicable regulatory authority in that market, and we may never receive such regulatory approval for any of our product candidates. To obtain separate regulatory approval in many other countries we must comply with numerous and varying regulatory requirements of such countries regarding safety and efficacy and governing, among other things, clinical trials, manufacturing and commercial sales, pricing and distribution of our product candidates, and we cannot predict success in these jurisdictions. If we obtain approval of our product candidates and ultimately commercialize our product candidates in markets outside of the United States, we would be subject to additional risks and uncertainties, including:

- our ability to obtain reimbursement for our product candidates in such markets;
- our inability to directly control commercial activities because we 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, which will last until December 31, 2020. During the transition period, most EU rules and regulations will continue to apply to us in the United Kingdom, and complex negotiations with the European Union relating to the future trading relationship between the parties will be ongoing. At this stage, the nature of the future relationship between the United Kingdom and the remaining European Union countries following the transition period has yet to be agreed, and ongoing negotiations with the European Union have demonstrated the difficulties that exist in reaching such an agreement. Significant political and economic uncertainty remains about whether the terms of the relationship will differ materially from the terms before withdrawal, and while the government of the United Kingdom has stated that it will negotiate the terms of a future trading arrangement with the European Union, there is no guarantee that the terms of such arrangement will be agreed or ratified by the government of the United Kingdom or the European Union prior to the scheduled 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. There are also risks arising from uncertainty in the regulatory environment including how clinical trials may be regulated. Although it is not possible to predict fully the effects of the United Kingdom's withdrawal from the European Union, any of these risks 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. For example, we announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY of rilonacept in recurrent pericarditis. Upon approval from the FDA, if any, of an sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. As we assume the sales and distribution responsibilities of rilonacept for the approved indications in the United States and begin commercializing rilonacept for the treatment of recurrent pericarditis or any of our other product candidates, if approved, we will be subject to additional ongoing obligations and continued regulatory review, which may result in significant additional expense.

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

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

While our current clinical and medical affairs activities are subject to certain ongoing regulatory requirements concerning appropriate exchange of medical and scientific information, 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. Further, the outcome of the upcoming U.S. presidential election may change governmental policies and regulations that affect our operations and business, including our clinical trials, regulatory approval, pharmaceutical pricing and reimbursement. 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 in the market, we announced positive data from our global, pivotal Phase 3 clinical trial, RHAPSODY, of rilonacept in recurrent pericarditis. Upon receipt of FDA approval, if any, of an sBLA submission for the commercial marketing of rilonacept in the United States for recurrent pericarditis, we will assume the sales and distribution of rilonacept for the approved indications in the United States and would evenly split profits on sales with Regeneron, after deducting certain commercialization expenses subject to specified limits. As we assume the sales and distribution responsibilities of rilonacept for the approved indications in the United States and begin commercializing rilonacept for the treatment or recurrent pericarditis or any of our other 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 or the easing of such measures, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position.

The COVID-19 pandemic, and measures taken in response to the pandemic or the easing of such measures, could cause significant disruption in our business and operations and could cause significant disruption the business and operations of our manufacturers, CROs upon whom we rely to conduct our clinical trials, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities. For example, our CRO for the Phase 1 clinical trial of KPL-404 notified us that, after a temporary pause due to the impact of the COVID-19 pandemic, they resumed certain clinical trial activities. While we do not expect this development will impact our timeline to report data from the trial in the second half of 2020, it will have an impact on the commencement of certain planned cohorts in the trial and the breadth of data we may be able to generate at that time should we conduct such other cohorts in the trial at another clinical trial site.

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 were and are considered to be an essential business so, to date, our operations have only been partially affected by these orders. In response to these orders, we implemented work-place rules and temporarily closed access to our California office space and restricted access to our Massachusetts facility to only those employees that needed to be in the office to execute their responsibilities and those employees who worked in our research and development laboratory space, with most of our employees continuing to carry out their responsibilities working outside of our offices. Subsequently in May and April 2020, the governors of Massachusetts and California, respectively, each announced a phased reopening plan for businesses and other organizations in their respective states. In response, we updated our work-place rules and designed a plan to reopen the Lexington and San Diego office spaces to additional groups of employees, in phases on an optional basis for now. Most of our employees continue to carry out their responsibilities working outside of our offices. We continue to monitor the developments, restrictions and requirements in jurisdictions where we have offices, and plan to update the protocols for our offices as applicable.

If the COVID-19 pandemic and measures undertaken in response to the pandemic are prolonged, or the easing of any of such measures has significant adverse consequences, we may experience and our manufacturers, CROs or other third parties with whom we conduct business or otherwise engage, may experience or continue to experience staffing shortages or reprioritizations, production slowdowns or stoppages, and disruptions in delivery systems now or in

the future. For example, the COVID-19 pandemic and measures taken in response to the pandemic, including business and travel restrictions and social-distancing to halt the spread of the pandemic, has had an impact on certain aspects of our commercialization strategy, including interacting with third-party payors, physicians and patient advocacy groups to build disease awareness, and conducting in-person market research as well as recruiting qualified candidates to enhance our commercial operations and support commercialization, which, if prolonged, may impede the effective commercialization of our product candidates and result in lower than anticipated future revenue.

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

Moreover, the COVID-19 pandemic is impacting the global economy, and the U.S. economy in particular, with the potential for the economic downturn to be severe and prolonged. A severe or prolonged economic downturn could result in a variety of risks to our business, including disruptions in the financial markets. For example, the trading prices of biopharmaceutical companies have been highly volatile as a result of the COVID-19 pandemic. These disruptions could adversely impact our ability to raise additional capital when needed or on acceptable terms, if at all.

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

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

We face an inherent risk of product liability exposure related to the testing of our product candidates in human clinical trials and 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. On March 2, 2020, the U.S. Supreme Court granted the petitions for writs of certiorari to review the case, although it is unclear when a decision will be made or how the Supreme Court will rule. In addition, there may be other efforts to challenge, repeal or replace the ACA, or portions thereof, which will affect our business. It is possible that the ACA, as currently enacted or as it may be amended in the future, and other healthcare reform measures that may be adopted in the future, could have a material adverse effect on our industry generally and on our ability to successfully commercialize our product candidates, if approved.

In addition, other legislative changes have been proposed and adopted in the United States since the Affordable Care Act was enacted. The Budget Control Act of 2011, among other things, led to aggregate reductions of Medicare payments to providers of 2% per fiscal year. These reductions went into effect on April 1, 2013 and, due to subsequent legislative amendments to the statute, will remain in effect through 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. For example, the outcome of the upcoming U.S. presidential election may change governmental policies and regulations that affect our operations and business, including our clinical trials, regulatory approval, pharmaceutical pricing and reimbursement. If we or any third party we may engage are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we or such third party are not able to maintain regulatory compliance, our product candidates may lose any regulatory approval that may have been obtained and we may not achieve or sustain profitability.

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

Our results of operations could be adversely affected by general conditions in the global economy and in the global financial markets. For example, the COVID-19 pandemic is impacting the global economy with the potential for the economic downturn to be severe and prolonged. A severe or prolonged economic downturn caused by the economic impact from the COVID-19 pandemic could result in a variety of risks to our business, such as disruptions to our operations and the operations of our manufacturers, CROs or other third parties with whom we conduct business or engage, including as a result of disruptions in travel into and within the countries in which we conduct our clinical trials or our manufacturers produce our product candidates or we conduct business or otherwise engage with such other third parties. These disruptions could adversely affect our ability to satisfy the required supply for any of our product candidates or successfully complete preclinical and clinical development of our product candidates, which could require us to incur additional costs, and impair our ability to obtain regulatory approval of our product candidates and generate revenue. A severe or prolonged economic downturn could also impair our ability to raise additional capital when needed

or on acceptable terms, if at all. Doing business internationally involves a number of other risks, including but not limited to:

- multiple, conflicting and changing laws and regulations such as privacy regulations, tax laws, export and import restrictions;
- employment laws, regulatory requirements and other governmental approvals, permits and licenses;
- failure by us to obtain and maintain regulatory approvals for the use of our products in various countries;
- additional potentially relevant third-party patent rights;
- complexities and difficulties in obtaining protection and enforcing our intellectual property;
- difficulties in staffing and managing operations outside of the United States;
- complexities associated with managing multiple payor reimbursement regimes, government payors or patient self-pay systems;
- limits in our ability to penetrate international markets;
- financial risks, such as longer payment cycles, difficulty collecting accounts receivable, the impact of local and regional financial crises on demand and payment for our products and exposure to foreign currency exchange rate fluctuations;
- natural disasters, political and economic instability, including wars, terrorism, political unrest, outbreak of disease and boycotts;
- curtailment of trade, and other business restrictions;
- certain expenses including, among others, expenses for travel, translation and insurance; and
- regulatory and compliance risks that relate to maintaining accurate information and control over sales and activities that may fall within the purview of the U.S. Foreign Corrupt Practices Act, its books and records provisions or its anti-bribery provisions.

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

Our internal technology systems, or those of our third-party CMOs, CROs or other contractors, consultants and service providers, may fail or suffer cyber-attacks or security breaches, which could result in a material disruption of our or such third-party's business or operations and our development programs for our product candidates' or loss of other assets, including funds.

Despite the implementation of security measures and cyber-security insurance, our internal technology systems and those of our third-party CMOs, CROs and other contractors, consultants and service providers as well as employees that are working outside of our facilities are vulnerable to damage from viruses, unauthorized access and attacks, theft, natural disasters, terrorism, war and telecommunication and electrical failures. As a result of the COVID-19 pandemic, we may experience increased cybersecurity risks due to the impacts from prolonged remote work arrangements. If such an event were to occur and cause interruptions in our business and operations or those of our third-party CMOs, CROs and other contractors, consultants and service providers as well as employees that are working outside of our facilities, it could result in a material disruption of our or such third-party's business or operations and our development programs of our product candidates' or loss of other assets, including funds. For example, the loss of clinical trial data for our product

candidates could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any disruption or security breach results in a loss of or damage to our data or applications or other data or applications relating to our technology or product candidates, or inappropriate disclosure or theft of confidential or proprietary information, we could incur liabilities not covered by our cyber-security insurance and the further development of our product candidates could be delayed.

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 General Data Protection Regulation 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, took effect in May 2018 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 European Union 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) 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 certain of our directors, and Class B1 common shares, all of which shares are held by entities affiliated with certain of our directors means that such persons are, and such entities may in the future be, able to influence certain matters submitted to our shareholders for approval; and such concentration and conversion rights and resulting concentration of control may have an adverse effect on the price of our Class A common shares and may result in our Class A common shares being undervalued.

Each Class A common share is entitled to one vote per Class A common share and each Class B common share is entitled to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares have no voting rights. As a result, all matters submitted to our shareholders are decided by the vote of holders of our Class A common shares and Class B common shares. As a result of the multi-class voting structure of our common shares, the

holders of our Class B common shares, which consist primarily of our executive officers and certain other members of our senior management, collectively control over a majority of the combined voting power of our common shares and therefore are able to control the outcome of certain matters submitted to our shareholders for approval. As of September 30, 2020, the holders of Class A common shares accounted for approximately 57% of our aggregate voting power and the holders of Class B common shares accounted for approximately 43% 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 39% of our aggregate voting power as of September 30, 2020 and have the ability to influence the outcome of certain matters submitted to our shareholders for approval.

However, this percentage may change depending on any conversion of our Class B common shares, Class A1 common shares or Class B1 common shares. Each holder of our Class B common shares has the ability to convert any portion of its Class B common shares into Class B1 common shares or Class A common shares at any time with advance notice to us. Each holder of our Class B1 common shares has the ability to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time with advance notice to us, and each holder of our Class A1 common shares has the ability to convert any portion of its Class A1 common shares into Class A common shares at any time with advance notice to us. Our Class A1 common shares and Class B1 common shares cannot be converted if, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of our issued and outstanding Class A common shares unless such holder provides us with 61-days' prior notice that it intends to increase, decrease or waive such threshold upon conversion. As of September 30, 2020, entities affiliated with certain members of our directors could convert their Class A1 common shares and Class B1 common shares upon 61-days' prior written notice into Class A common shares and Class B common shares, respectively, which in the aggregate would result in such entities holding approximately 77% of our aggregate voting power and having the ability to control the outcome of certain matters submitted to our shareholders for approval.

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

These conversion rights as well as concentrated control that limit certain shareholders' ability to influence corporate matters may have an adverse effect on the price of our Class A common shares, including our Class A common shares being undervalued. Holders of our Class B common shares collectively control our management and affairs and are able to influence or control the outcome of certain matters submitted to our shareholders for approval, including the election of directors. Due to the conversion rights of the holders of our Class A1 and B1 common shares, entities affiliated with certain of our directors could significantly increase their voting control of us, which could result in their ability to significantly influence or control matters submitted to our shareholders for approval. As of September 30, 2020, entities affiliated with certain of our directors held 71% of our Class A1 common shares and 100% of our Class B1 common shares. Upon 61-days' prior written notice, these entities could convert their Class A1 common shares and Class B1 common shares into Class A common shares and Class B common shares, which in the aggregate would result in such entities holding approximately 77% of the voting power of our outstanding share capital. In addition, this concentration of control might adversely affect certain corporate actions that some of our shareholders may view as beneficial, for example, by:

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

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

Our share price is likely to continue to be volatile. The stock market in general and the market for biopharmaceutical companies in particular have experienced extreme volatility, including as a result of the COVID-19 pandemic, that has often been unrelated to the operating performance of particular companies. As a result of this volatility, our shareholders may not be able to sell their Class A common shares at or above the price they paid for their shares. The market price for our Class A common shares may be influenced by many factors, including:

- the results of clinical trials for our product candidates;
- delays in the commencement, enrollment and the ultimate completion of clinical trials;
- the results and potential impact of competitive products or technologies;
- our ability to manufacture and successfully produce our product candidates;
- our ability to commercialize our product candidates, if approved;
- the size of the market for our product candidates;
- actual or anticipated changes in estimates as to financial results, capitalization, development timelines or recommendations by securities analysts;
- the level of expenses related to any of our product candidates or clinical development programs;
- variations in our financial results or those of companies that are perceived to be similar to us;
- financing or other corporate transactions, or our inability to obtain additional funding;
- failure to meet or exceed the expectations of the investment community;
- regulatory or legal developments in the United States and other countries;
- the U.S. presidential election;
- the recruitment or departure of key personnel;
- developments or disputes concerning patent applications, issued patents or other proprietary rights;
- the results of our efforts to discover, develop, acquire or in-license additional product candidates or from our entering into entering collaborations or other strategic transaction agreements;
- changes in the structure of healthcare payment systems;
- market conditions in the pharmaceutical and biotechnology sectors;
- general economic, industry and market conditions, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic;
- changes in voting control of, or sales of our shares by, our executive officers and certain other members of our senior management or entities affiliated with certain of our directors that hold our shares; and

- the other factors described in this “Risk Factors” section.

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

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

If securities or industry analysts cease publishing or publish unfavorable research or reports about us, our business or our market, our shares price and trading volume could decline.

The trading market for our Class A common shares is influenced by the research and reports that equity research analysts publish about us and our business. We do not have any control over the analysts or the content and opinions included in their reports. The price of our Class A common shares could decline if one or more equity research analysts downgrades our shares or issues other unfavorable commentary or research. If one or more equity research analysts ceases coverage of our company or fails to publish reports on us regularly, demand for our Class A common shares could decrease, which in turn could cause the price of our Class A common shares or its trading volume to decline.

Sales of a number of our Class A common shares in the public market, including Class A common shares issuable upon conversion of our Class B, Class A1 and Class B1 common shares, could cause the share price of our Class A common shares to fall.

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

As of September 30, 2020, upon conversions of our Class B, Class A1, and Class B1 common shares, approximately 2.3 million of additional Class A common shares were issuable and eligible for resale in the public market to the extent permitted by the provisions of Rule 144 promulgated under the Securities Act of 1933, as amended, or the Securities Act, and such rule, Rule 144. In addition, as of September 30, 2020, there were approximately 10.4 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 entities affiliated with certain of our directors. As of September 30, 2020, on an as-converted to Class A common shares basis, these shareholders collectively held approximately 34.5 million of our Class A common shares. If any of these shareholders sell, convert or transfer, or indicate an intention to sell, convert or transfer, a substantial amount of their common shares (after certain restrictions on conversion or resale lapse), the market price of our Class A common shares could decline.

Pursuant to our amended and restated investor rights agreement, or our investors rights agreement, certain shareholders are entitled to certain registration rights with respect our Class A common shares, including Class A common shares issuable upon conversions of our Class B, Class A1, and Class B1 common shares and upon the exercise of certain rights to acquire Class A common shares, or collectively registerable securities, under the Securities Act. As of September 30, 2020, on an as-converted to Class A common shares basis, we have registered approximately 31.8 million Class A common shares held by certain holders affiliated with certain of our directors as well as certain other shareholders pursuant to our investor rights agreement, which are freely tradable without restriction under the Securities Act, to the extent permitted by Rule 144. Further, pursuant to the investors rights agreement (a) the holders affiliated with certain of our directors are entitled to certain registration rights under the Securities Act with respect to registrable securities they may own now or in the future and (b) our executive officers are also entitled to certain registration rights under the Securities Act with respect to registrable securities they may own now or in the future, including, on an as-converted to Class A common shares basis, the approximately 1.9 million Class A common shares held by our executive officers as of September 30, 2020. 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 (a) upfront consideration of $10.0 million paid at closing in March 2019 as well as milestone payments of $5.0 million that had been achieved and was also paid at closing and (b) a milestone payment of $3.0 million that had been achieved after the closing and 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 share amounts) in accordance with the terms and conditions of our stock purchase option agreement with Primatope, or the Primatope Agreement. In June 2020, we released the escrow and in June and September of 2020 we issued all of the shares held back in connection with the acquisition.

We are currently 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 currently 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.

Based on our closing share price and the market value of our voting and non-voting common shares held by non-affiliates as of June 30, 2020, we have determined that we will no longer be an emerging growth company as of December 31, 2020. However, 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;
- simplified executive compensation disclosure; 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 based on our closing share price and the market value of our voting and non-voting common shares held by non-affiliates as of June 30, 2020, we have determined that we will no longer be a smaller reporting company as of January 1, 2021. However, for so long as we remain a smaller reporting company, we are permitted and intend to rely on exemptions from certain disclosure requirements that are applicable to other public companies that are not smaller reporting companies. 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, including in connection with becoming a large accelerated filer as of December 31, 2020, and our management will be required to devote substantial time to new compliance initiatives.

As a public company, we incur significant legal, accounting and other expenses. In addition, the Sarbanes-Oxley Act of 2002 and rules subsequently implemented by the SEC and The Nasdaq Global Select Market, or Nasdaq, where our Class A common shares are listed, impose various requirements on public companies, including

establishment and maintenance of effective disclosure and financial controls and corporate governance practices. Our management and other personnel devote a substantial amount of time to these compliance initiatives. Moreover, these rules and regulations increase our legal and financial compliance costs and make some activities more time-consuming and costly, which will increase in connection with our becoming a large accelerated filer and no longer being an emerging growth company and 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. In addition, we expect our costs to increase as we comply with requirements related to executive compensation disclosure and shareholder votes on executive compensation matters that we were previously exempt from as an emerging growth company and smaller reporting company. While we are currently an emerging growth company and a smaller reporting company, based on our closing share price and the market value of our voting and non-voting common shares held by non-affiliates as of June 30, 2020, we have determined that we will be a large accelerated file and no longer be an emerging growth company as of December 31, 2020 and no longer be a smaller reporting company as of January 1, 2020. However, for so long as we remain an emerging growth company and smaller reporting 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.

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. As an emerging growth company, we were not required to include an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. However, based on our determination that we will be a large accelerated filer and cease being an emerging growth company as of December 31, 2020, we will need to do so. 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. We expect these activities to increase in connection with our auditors' review of our internal control over financial reporting in preparation for providing their attestation report. Despite our efforts, there is a risk that neither we nor our independent registered public accounting firm will be able to conclude that our internal control over financial reporting is effective as required by Section 404. This could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statements.

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

Our amended and restated bye-laws contain provisions that could make it more difficult for a third party to acquire us. These provisions provide for:

- a classified board of directors with staggered three-year terms;
- directors only to be removed for cause;
- an affirmative vote of $66^{2}/_{3}\%$ of the voting power of our voting shares for certain "business combination" transactions that have not been approved by our board of directors;
- our multi-class common share structure, which provides our holders of Class B common shares with the ability to significantly influence the outcome of matters requiring shareholder approval, even if they own less than a majority of our outstanding Class A common shares;
- restrictions on the time period in which directors may be nominated; and
- our board of directors to determine the powers, preferences and rights of our preferred shares and to issue the preferred shares without shareholder approval.

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

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

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

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

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

We are a Bermuda exempted company. As a result, the rights of holders of our Class A common shares will be governed by Bermuda law and our memorandum of association and amended and restated bye-laws. The rights of shareholders under Bermuda law may differ from the rights of shareholders of companies incorporated in other jurisdictions. It may be difficult for investors to enforce in the United States judgments obtained in U.S. courts against us based on the civil liability provisions of the U.S. securities laws. It is doubtful whether courts in Bermuda will enforce judgments obtained in other jurisdictions, including the United States, against us or our directors or officers under the securities laws of those jurisdictions or entertain actions in Bermuda against us or our directors or officers under the securities laws of other jurisdictions.

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

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

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

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

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

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

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

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

We may become subject to unanticipated tax liabilities.

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

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

We are incorporated under the laws of Bermuda and currently have subsidiaries in the United States, the United Kingdom, Germany, Switzerland and France. If we succeed in growing our business, we expect to conduct increased operations through our subsidiaries in various tax jurisdictions subject to transfer pricing arrangements between us and such subsidiaries. If two or more affiliated companies are located in different countries, the tax laws or regulations of

each country generally will require that transfer prices be the same as those between unrelated companies dealing at arms' length and that appropriate documentation is maintained to support the transfer prices. While we believe that we operate in compliance with applicable transfer pricing laws and intend to continue to do so, our transfer pricing procedures are not binding on applicable tax authorities.

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

Changes in laws related to tax practices and substance requirements in Bermuda and other jurisdictions could adversely affect our operations.

Our tax position could be adversely impacted by changes in tax rates, tax laws, tax practice, tax treaties or tax regulations or changes in the interpretation thereof by the tax authorities in Europe (including the United Kingdom), the United States, Bermuda and other jurisdictions, as well as being affected by certain changes currently proposed by the Organization for Economic Co-operation and Development and their action plan on Base Erosion and Profit Shifting. Such changes may become more likely as a result of recent economic trends in the jurisdictions in which we operate, particularly if such trends continue. If such a situation 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, which was amended as recently as December 24, 2019, or the Substance Act, and issued Economic Substance Regulations in 2018, which were amended as recently as December 24, 2019). Pursuant to the Substance Act and Economic Substance Regulations, certain entities in Bermuda engaged in "relevant activities" are required to maintain

physical presence in Bermuda and to satisfy economic substance requirements commencing as of January 1, 2019, with a six-month transition period until July 1, 2019. The list of "relevant activities" includes carrying on as a business in any one or more of the following categories: banking, insurance, fund management, financing, leasing, headquarters, shipping, distribution and service center, intellectual property and holding entities. Under the Substance Act, any relevant entity must satisfy economic substance requirements locally or face financial penalties, restriction or regulation of its business activities or may be struck off as a registered entity from the Bermuda Registrar of Companies. Because we do not report gross revenue attributable to any such relevant activity, under Section 6 of the Revised Final Guidance Notes issued on September 18,2020, we believe that we are not obliged to meet the economic substance requirements. We will continue to monitor our status with respect to the 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. We believe that our subsidiaries based in the United Kingdom, Germany, Switzerland and France, will not be classified as a PFIC for the 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

Issuance of Unregistered Equity Securities

On May 18, 2020, we issued and sold an aggregate of 1,600,000 Class A1 common shares to existing investors at price of $18.25 per share, resulting in aggregate gross proceeds to us of approximately $29.2 million. These securities were issued under Section 4(a)(2) of the Securities Act in a transaction not involving a public offering.

On June 6, 2020, we issued an aggregate of 59,469 Class A common shares to the former shareholders of Primatope, having an aggregate value of approximately less than $0.1 million, in connection with the release and issuance of the shares held back at the closing of the acquisition of Primatope, in accordance with the Primatope Agreement. These securities were issued under Section 4(a)(2) and Rule 506 of the Securities Act in a transaction not involving a public offering.

On July 24, 2020, we issued and sold an aggregate of 1,428,572 Class A1 common shares to existing investors at price of $21.00 per share, resulting in aggregate gross proceeds to us of approximately $30.0 million. These securities were issued under Section 4(a)(2) of the Securities Act in a transaction not involving a public offering.

On September 4, 2020, we issued an aggregate of 16,634 Class A common shares to the former shareholders of Primatope, having an aggregate value of approximately $0.3 million, as payment, in part, for the achievement a milestone in accordance with the Primatope Agreement. These securities were issued under Section 4(a)(2) and Rule 506 of the Securities Act in a transaction not involving a public offering.

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	Memorandum of Association of Kiniksa Pharmaceuticals, Ltd.	S-1	333-224488	3.1	4/27/2018	
3.2	Amended and Restated Bye-Laws of Kiniksa Pharmaceuticals, Ltd.	8-K	001-38492	3.1	5/29/2018	
4.1	Specimen Share Certificate evidencing the Class A common shares	S-1/A	333-224488	4.1	5/14/2018	
4.2	Second Amended and Restated Investors' Rights Agreement, dated as of February 9, 2018	S-1	333-224488	3.1	4/27/2018	
31.1	Rule 13a-14(a) / 15d-14(a) Certification of Principal Executive Officer					*
31.2	Rule 13a-14(a) / 15d-14(a) Certification of Principal Financial Officer					*
32.1	Section 1350 Certification of Principal Executive Officer					**
32.2	Section 1350 Certification of Principal Financial Officer					**
101.INS	XBRL Instance Document – the instance document does not appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL document					***
101.SCH	Inline XBRL Taxonomy Extension Schema Document					***
101.CAL	Inline XBRL Taxonomy Extension Calculation Linkbase Document					***
101.DEF	Inline XBRL Extension Definition Linkbase Document					***
101.LAB	Inline XBRL Taxonomy Label Linkbase Document					***
101.PRE	Inline XBRL Taxonomy Extension Presentation Linkbase Document					***
104	Cover page Interactive Data File (formatted in Inline XBRL and contained in Exhibit 101) - The cover page interactive data file does not					***

Exhibit Number	Exhibit Description	Incorporated by Reference				Filed/ Furnished Herewith
		Form	File No.	Exhibit	Filing Date	
	appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL 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: November 5, 2020

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

Exhibit 31.1

CERTIFICATIONS

I, Sanj K. Patel, certify that:

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

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

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

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

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

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

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

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

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

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

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

November 5, 2020

/s/ Sanj K. Patel

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

Exhibit 31.2

CERTIFICATIONS

I, 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.

November 5, 2020

/s/ Michael R. Megna

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

Exhibit 32.1

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

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

(1) The Quarterly Report on Form 10-Q of the Company for the period ended September 30, 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.

November 5, 2020

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

Exhibit 32.2

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

I, 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 September 30, 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.

November 5, 2020

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