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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 June 30, 2019
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 Number)

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)

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 ☒

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

As of July 31, 2019, there were 54,857,692 common shares outstanding in aggregate, comprised of:

19,165,265 Class A common shares, par value $0.000273235 per share

4,638,855 Class B common shares, par value $0.000273235 per share

14,995,954 Class A1 common shares, par value $0.000273235 per share

16,057,618 Class B1 common shares, par value $0.000273235 per share

Kiniksa Pharmaceuticals, Ltd.

FORM 10-Q

FOR THE THREE MONTHS ENDED JUNE 30, 2019

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SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This Quarterly Report on Form 10-Q, or Quarterly Report, contains forward-looking statements. All statements other than statements of historical facts contained in this Quarterly Report including statements regarding our future results of operations and financial position, business strategy, prospective products and product candidates, their expected properties, performance and impact on healthcare costs, the expected timeline for achievement of our clinical milestones, the timing of, and potential results from, clinical and other trials, marketing authorization from the FDA or regulatory authorities in other jurisdictions, coverage and reimbursement for procedures using our product candidates, if approved, research and development costs, timing of regulatory filings and feedback, timing and likelihood of success, plans and objectives of management for future operations and future results of anticipated products, are forward-looking statements.

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

In some cases, you can identify forward-looking statements by terms such as “may,” “will,” “should,” “expect,” “plan,” “anticipate,” “could,” “intend,” “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, including, without limitation, the following:

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

- the size of the market for our product candidates;
- our ability to meet the quality expectations of physicians or patients;
- our ability to improve our product candidates;
- the decision of third-party payors not to cover our product candidates or to require extensive or independently performed clinical trials prior to covering or maintaining coverage of our product candidates;
- our ability to successfully manage our growth;
- our ability to avoid product liability claims and maintain adequate product liability insurance;
- our ability to obtain regulatory exclusivity;
- our ability to obtain, maintain, protect and enforce our intellectual property rights related to our product candidates;
- federal, state and foreign regulatory requirements applicable to our product candidates; and
- ownership concentration of our executive officers and certain members of senior management may prevent our shareholders from influencing significant corporate decisions.

Because forward-looking statements are inherently subject to risks and uncertainties, some of which cannot be predicted or quantified and some of which are beyond our control, you should not rely on these forward-looking statements as predictions of future events. The events and circumstances reflected in our forward-looking statements may not be achieved or occur and actual results could differ materially from those projected in the forward-looking statements. Moreover, we operate in an evolving environment. New risk factors and uncertainties may emerge from time to time, and it is not possible for management to predict all risk factors and uncertainties. As a result of these factors, we cannot assure you that the forward-looking statements in this 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.

Part I — Financial Information

Item 1. Financial Statements (unaudited)

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

	June 30, 2019	December 31, 2018
Assets		
Current assets:		
Cash and cash equivalents	$ 82,810	$ 71,976
Short-term investments	204,637	235,328
Prepaid expenses and other current assets	9,011	6,446
Total current assets	296,458	313,750
Property and equipment, net	6,359	6,356
Operating lease right-of-use assets	2,503	—
Restricted cash	210	210
Deferred offering costs	—	433
Deferred tax assets	2,607	1,216
Total assets	$ 308,137	$ 321,965
Liabilities and Shareholders' Equity		
Current liabilities:		
Accounts payable	$ 9,338	$ 10,918
Accrued expenses	19,179	16,418
Accrued milestones	—	15,000
Operating lease liabilities	1,496	—
Other current liabilities	165	218
Total current liabilities	30,178	42,554
Noncurrent liabilities:		
Noncurrent operating lease liabilities	1,819	—
Other long-term liabilities	943	144
Total liabilities	32,940	42,698
Commitments and contingencies (Note 12)		
Shareholders' equity:		
Class A common shares, par value of $0.000273235 per share; 19,165,265 shares and 15,797,220 shares issued and outstanding as of June 30, 2019 and December 31, 2018, respectively	6	4
Class B common shares, par value of $0.000273235 per share; 4,638,855 shares issued and outstanding as of June 30, 2019 and December 31, 2018	1	1
Class A1 common shares, $0.000273235 par value; 14,995,954 shares and 12,995,954 shares issued and outstanding as of June 30, 2019 and December 31, 2018, respectively	4	4
Class B1 common shares, $0.000273235 par value; 16,057,618 shares issued and outstanding as of June 30, 2019 and December 31, 2018	4	4
Additional paid-in capital	572,318	473,483
Accumulated other comprehensive income (loss)	101	(4)
Accumulated deficit	(297,237)	(194,225)
Total shareholders' equity	275,197	279,267
Total liabilities and shareholders' equity	$ 308,137	$ 321,965

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 June 30,		Six Months Ended June 30,	
	2019	2018	2019	2018
Operating expenses:				
Research and development	$ 30,848	17,200	$ 90,101	29,831
General and administrative	8,441	4,327	16,835	8,036
Total operating expenses	39,289	21,527	106,936	37,867
Loss from operations	(39,289)	(21,527)	(106,936)	(37,867)
Interest income	1,724	1,066	3,533	1,371
Loss before benefit for income taxes	(37,565)	(20,461)	(103,403)	(36,496)
Benefit for income taxes	374	202	391	255
Net loss	$ (37,191)	$ (20,259)	$ (103,012)	$ (36,241)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.68)	$ (1.11)	$ (1.94)	$ (3.45)
Weighted average common shares outstanding—basic and diluted	54,475,476	18,328,402	53,225,710	10,492,474
Comprehensive loss:				
Net loss	$ (37,191)	$ (20,259)	$ (103,012)	$ (36,241)
Other comprehensive income:				
Unrealized gain on short-term investments	93	—	105	—
Total other comprehensive income	93	—	105	—
Total comprehensive loss	$ (37,098)	$ (20,259)	$ (102,907)	$ (36,241)

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

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

	Convertible Preferred Shares (Series A, B and C)		Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income (Loss)	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount	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 offering costs	—	—	2,816,110	2	48,474	—	—	48,476
Issuance of Class A1 common shares upon completion of private placement, net of underwriting discounts and commissions and offering costs	—	—	2,000,000	—	34,511	—	—	34,511
Class A common shares issued or to be issued in connection with the acquisition of all issued and outstanding equity securities of Primatope Therapeutics, Inc.	—	—	337,008	—	7,000	—	—	7,000
Exercise of options	—	—	50,070		181	—	—	181
Share-based compensation expense	—	—	—	—	2,893	—	—	2,893
Unrealized gain on short-term investments	—	—	—	—	—	12	—	12
Net loss	—	—	—	—	—	—	(65,821)	(65,821)
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

	Convertible Preferred Shares (Series A, B and C)		Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount	Shares	Amount				
Balances at December 31, 2017	22,885,492	119,770	4,288,329	$ 1	$ 1,289	$ —	$ (90,998)	$ (89,708)
Issuance of Series C convertible preferred shares, net of issuance costs of $9,178	12,784,601	190,822	—	—	—	—	—	—
Exercise of options	—	—	4,574	—	17	—	—	17
Share-based compensation expense	—	—	—	—	558	—	—	558
Net loss	—	—	—	—	—	—	(15,982)	(15,982)
Balances at March 31, 2018	35,670,093	310,592	4,292,903	$ 1	$ 1,864	$ —	$ (106,980)	$ (105,115)
Conversion of convertible preferred shares to common shares	(35,670,093)	(310,592)	35,670,093	8	310,584	—	—	310,592
Issuance of Class A common shares upon completion of initial public offering, net of underwriting discounts and commissions and offering costs	—	—	9,484,202	4	155,511	—	—	155,515
Share-based compensation expense	—	—	—	—	1,059	—	—	1,059
Unrealized gain on short term investments	—	—	—	—	—	7	—	7
Net loss	—	—	—	—	—	—	(20,259)	(20,259)
Balances at June 30, 2018	—	—	49,447,198	$ 13	$ 469,018	$ 7	$ (127,239)	$ 341,799

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

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

	Six Months Ended June 30,	
	2019	**2018**
Cash flows from operating activities:		
Net loss	$ (103,012)	$ (36,241)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	979	20
Share-based compensation expense	6,357	1,617
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	—	66
Non-cash rent expense	—	208
Accretion of discounts on short-term investments	(2,055)	—
Deferred income taxes	(1,390)	(463)
Changes in operating assets and liabilities:		
Prepaid expenses, right-of-use assets and other assets	(1,009)	(2,965)
Accounts payable	(1,663)	(747)
Accrued expenses and other liabilities	2,720	5,616
Accrued milestones	(15,000)	—
Operating lease liabilities	(602)	—
Other long-term liabilities	943	—
Net cash used in operating activities	(104,932)	(32,889)
Cash flows from investing activities:		
Purchases of property and equipment	(767)	(311)
Purchases of short-term investments	(273,488)	(73,676)
Proceeds from the maturities of short-term investments	306,340	—
Net cash provided by (used in) investing activities	32,085	(73,987)
Cash flows from financing activities:		
Proceeds from issuance of Series C convertible preferred shares, net of issuance costs	—	190,822
Proceeds from issuance of Class A common shares upon completion of initial public offering, net of underwriting commissions and discounts, inclusive of the over-allotment option exercise	—	159,193
Proceeds from issuance of Class A common shares from follow-on offering, net of underwriting commissions and discounts, inclusive of the over-allotment option exercise	48,595	—
Proceeds from issuance of Class A1 common shares from private placement, net of underwriting commissions and discounts	34,511	—
Payments of offering costs	(118)	(2,825)
Proceeds from exercise of options and employee share purchase plan	693	17
Net cash provided by financing activities	83,681	347,207
Net increase in cash and cash equivalents and restricted cash	10,834	240,331
Cash and cash equivalents and restricted cash at beginning of period	72,186	45,660
Cash and cash equivalents and restricted cash at end of period	$ 83,020	$ 285,991
Supplemental information:		
Cash paid for income taxes	$ 1,027	$ 148
Supplemental disclosure of non-cash investing and financing activities:		
Deferred offering costs included in accrued expenses and accounts payable	$ —	$ 856
Property and equipment included in accrued expenses and accounts payable	$ 253	$ —

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, focused on autoinflammatory and autoimmune conditions.

The Company is subject to risks and uncertainties common to early-stage companies in the biopharmaceutical industry. There can be no assurance that the Company's research and development will be successfully completed, that adequate protection for the Company's technology will be obtained, that any products developed will obtain necessary government regulatory approval or that any products, if approved, will be commercially viable. The Company does not currently generate revenue from sales of any products, and it may never be able to develop or commercialize a marketable product. The Company has not yet successfully completed any Phase 3 or other pivotal clinical trials, obtained any regulatory approvals, manufactured a commercial-scale drug, or conducted sales and marketing activities. The Company operates in an environment of rapid technological innovation and substantial competition from pharmaceutical and biotechnology companies. In addition, the Company is dependent upon the services of its employees, consultants and service providers. Even if the Company's product development efforts are successful, it is uncertain when, if ever, the Company will realize significant revenue from product sales.

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"), Kiniksa Pharmaceuticals (UK), Ltd. ("Kiniksa UK"), Kiniksa Pharmaceuticals (Germany) GmbH ("Kiniksa Germany"), Kiniksa Pharmaceuticals (France) SARL ("Kiniksa France") and Primatope Therapeutics, Inc. ("Primatope"), after elimination of all significant intercompany accounts and transactions.

In assessing the consolidation requirement for variable interest entities ("VIEs"), the Company focuses on identifying whether it has both the power to direct the activities that most significantly impact the VIE's economic performance and the obligation to absorb losses or the right to receive benefits from the VIE. In the event that the Company is the primary beneficiary of a VIE, the assets, liabilities, and results of operations of the VIE would be included in the Company's consolidated financial statements. At December 31, 2018 and during the year then ended and at June 30, 2019 and during the three and six months 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.

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

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 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 June 30, 2019 and the results of its operations for the three and six months ended June 30, 2019 and 2018 and its cash flows for the six months ended June 30, 2019 and 2018. The results for the three and six months ended June 30, 2019 are not necessarily indicative of results to be expected for the year ending December 31, 2019, any other interim periods or any future year or period.

Reverse Stock Split

On May 11, 2018, the Company effected a 1-for-2.73235 reverse share split of its authorized, designated, issued and outstanding common shares and preferred shares. Accordingly, all share and per share amounts for all periods presented in the accompanying consolidated financial statements and notes thereto have been adjusted retroactively, where applicable, to reflect this reverse share split.

Initial Public Offering

On May 23, 2018, the Company's registration statement on Form S-1 relating to its initial public offering of its Class A common shares (the "IPO") was declared effective by the Securities and Exchange Commission ("SEC"). On May 29, 2018, the Company completed the IPO of 8,477,777 Class A common shares at a public offering price of $18.00 per share for gross proceeds of $152,600. In addition, on June 22, 2018, the Company completed the sale of 1,006,425 Class A common shares to the underwriters of the IPO following the exercise in part of their over-allotment option to purchase additional shares at a public offering price of $18.00 per share for gross proceeds of $18,116. The aggregate net proceeds to the Company from the IPO, inclusive of the over-allotment option exercise, was $155,536 after deducting underwriting discounts and commissions and other offering costs.

Upon the closing of the IPO, all convertible preferred shares then outstanding automatically converted into 5,546,019 Class A common shares, 1,070,502 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares. In connection with the closing of the IPO, the Company amended and restated its bye-laws (the "Amended & Restated Bye-Laws").

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

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 $82,985 after deducting underwriting discounts and commissions and other offering costs

Liquidity

In accordance with Accounting Standards Update ("ASU") No. 2014-15, Disclosure of Uncertainties about an Entity's Ability to Continue as a Going Concern (Subtopic 205-40), the Company has evaluated whether there are conditions and events, considered in the aggregate, that raise substantial doubt about the Company's ability to continue as a going concern within one year after the date the consolidated financial statements are issued. As of June 30, 2019, the Company had an accumulated deficit of $297,237. During the six months ended June 30, 2019, the Company incurred a net loss of $103,012 and used $104,932 of net cash in operating activities. The Company expects to continue to generate operating losses for the foreseeable future. As of June 30, 2019, the Company had cash, cash equivalents and short-term investments of $287,447.

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 12 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. 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, 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 June 30, 2019 and December 31, 2018, cash and cash equivalents consisted principally of U.S. Treasury notes, amounts held in money market funds and cash on deposit at commercial banks.

Short-Term Investments

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

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

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.

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 June 30, 2019 and December 31, 2018, 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 June 30, 2019 and December 31, 2018, the underlying cash balance of $210 securing this letter of credit, was classified as non-current in its consolidated balance sheet.

Fair Value Measurements

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

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

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

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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 new standard on January 1, 2019 using the modified retrospective transition approach as applied to leases existing as of the adoption date. The standard will be 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 right-of-use asset being recorded on the balance sheet and amortized on a straight-line basis as lease expense.

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Research and Development Costs

Research and development costs are expensed as incurred. Research and development expenses consist of costs incurred to discover, research and develop drug candidates, including personnel expenses, share-based compensation expense, allocated facility-related and depreciation expenses, third-party license fees and external costs of outside vendors engaged to conduct preclinical and clinical development activities and clinical trials as well as to manufacture clinical trial materials. Non-refundable prepayments for goods or services that will be used or rendered for future research and development activities are recorded as prepaid expenses. Such amounts are recognized as an expense as the goods are delivered or the related services are performed, or until it is no longer expected that the goods will be delivered, or the services rendered.

Research Contract Costs and Accruals

The Company has entered into various research and development-related contracts with companies both inside and outside of the United States. The related costs are recorded as research and development expenses as incurred. The Company records accruals for estimated ongoing research costs. When evaluating the adequacy of the accrued liabilities, the Company analyzes progress of the studies or clinical trials, including the phase or completion of events, invoices received and contracted costs. Significant judgments and estimates are made in determining the accrued balances at the end of any reporting period. Actual results could differ materially from the Company's estimates. The Company's historical accrual estimates have not been materially different from the actual costs.

Share-Based Compensation

The Company measures all share-based awards granted to employees and directors based on their fair value on the date of the grant and recognizes compensation expense for those awards over the requisite service period, which is generally the vesting period of the respective award. Forfeitures are accounted for as they occur. The Company issues share-based awards with only service-based vesting conditions and records the expense for these awards using the straight-line method. The Company has not issued any share-based awards with performance-based vesting conditions.

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

The Company classifies share-based compensation expense in its consolidated statements of operations and comprehensive loss in the same manner in which the award recipient's payroll costs are classified or in which the award recipient's service payments are classified.

The fair value of each restricted share award is estimated on the date of grant based on the fair value of the Company’s Class A common shares or Class B common shares on that same date. The fair value of each option grant is estimated using the Black-Scholes option-pricing model, which requires inputs based on certain subjective assumptions, including the expected share price volatility, the expected term of the award, the risk-free interest rate, and expected dividends (see Note 9). Prior to May 2018, the Company was a private company and, accordingly, lacks company-specific historical and implied volatility information for its shares. Therefore, it estimates its expected share price volatility based on 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

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risk-free interest rate is determined by reference to the U.S. Treasury yield curve in effect at the time of grant of the award for time periods approximately equal to the expected term of the award. Expected dividend yield is based on the fact that the Company has never paid cash dividends on common shares and does not expect to pay any cash dividends in the foreseeable future.

Comprehensive Loss

Comprehensive loss includes net loss as well as other changes in shareholders' equity that result from transactions and economic events other than those with shareholders. For the three and six months ended June 30, 2019, the Company's other comprehensive loss was primarily related to unrealized gain on short-term investments as well as cumulative translation adjustments. For the three and six months ended June 30, 2018, there was no difference between net loss and comprehensive loss.

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 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, unvested restricted common shares and convertible preferred shares are considered potential dilutive common shares.

Prior to the closing of the IPO, when the Company's convertible preferred shares converted to common shares, the Company's convertible preferred shares contractually entitled the holders of such shares to participate in dividends but did not contractually require the holders of such shares to participate in losses of the Company. Accordingly, for periods in which the Company reported a net loss attributable to common shareholders, such losses were not allocated to convertible preferred shareholders. 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 six months ended June 30, 2019 and 2018.

The Company identified an error in its calculation of weighted average shares for certain shares issued and outstanding during the three and six months ended June 30, 2018, which is not considered material to the previously issued financial statements. The previously issued financial statements were revised to reflect an adjustment to decrease weighted average common shares outstanding by 2,458,886 and 1,243,104 and increase net loss per share by $0.14 and $0.36 for the three and six months ended June 30, 2018, respectively.

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Recently Adopted Accounting Pronouncements

In February 2016, the FASB issued ASU 2016-02, *Leases*. The standard, including subsequently issued amendments, collectively referred to as ASC 842, established principles of recognition, measurement, presentation and disclosure of lease arrangements applicable to lessees and lessors in order to enhance the transparency and compatibility of financial reporting related to the arrangements, including with respect to the amount, timing and uncertainty of cash flows arising from a lease. The Company adopted new accounting guidance regarding the accounting for leases as of January 1, 2019 using a modified retrospective transition approach that was applied to leases existing as of, or entered into prior to, January 1, 2019. See Note 2, Summary of Significant Accounting Policies, "Leases" for a discussion of the Company's policy with respect to this standard and Note 5, "Leases" for a discussion of the Company's adoption of this standard and its impact on its consolidated financial statements and related disclosures.

Upon the adoption of ASC 842, the Company recorded operating lease right-of-use assets of $3,682 and operating lease liabilities of $3,917 for its leases which were in effect and had commenced prior to January 1, 2019 and had original lease terms of more than 12 months.

3. Fair Value of Financial Assets and Liabilities

Short-term investments as of June 30, 2019 and December 31, 2018 consisted of U.S. Treasury notes which investments are each due within six months of such respective period. As of June 30, 2019 and December 31, 2018, the fair value of short-term investments was $204,637 and $235,328, respectively. As of June 30, 2019, the amortized cost was $204,536 and gross unrealized gain was $101. As of December 31, 2018, the amortized cost was $235,332 and gross unrealized loss was $4.

The following tables present information about the Company's financial assets 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 June 30, 2019 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 210	$ —	$ —	$ 210
Cash equivalents — money market funds	5,460	—	—	5,460
Cash equivalents — U.S. Treasury notes	—	15,485	—	15,485
Short-term investments — U.S. Treasury notes	—	204,637	—	204,637
	$ 5,670	$ 220,122	$ —	$ 225,792

	Fair Value Measurements as of December 31, 2018 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash — money market funds	$ 210	$ —	$ —	$ 210
Cash equivalents — money market funds	29,721	—	—	29,721
Cash equivalents — U.S. Treasury notes	—	15,634	—	15,634
Short-term investments — U.S. Treasury notes	—	235,328	—	235,328
	$ 29,931	$ 250,962	$ —	$ 280,893

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During the periods ended June 30, 2019 and December 31, 2018 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 June 30, 2019 and December 31, 2018 consisted of U.S. Treasury notes, which are not traded on a daily basis and, therefore, represent a Level 2 measurement in the fair value hierarchy at each period end. All of the Company's other assets and liabilities are recorded at fair value.

4. Property and Equipment, Net

Property and equipment, net consisted of the following:

	June 30, 2019	December 31, 2018
Furniture, fixtures and vehicles	$ 91	$ 91
Computer hardware and software	269	249
Leasehold improvements	3,225	2,676
Lab equipment	3,939	3,107
Construction in progress	133	552
Total property and equipment	7,657	6,675
Less: Accumulated depreciation	(1,298)	(319)
Total property and equipment, net	$ 6,359	$ 6,356

As of June 30, 2019, construction in progress is primarily comprised of lab equipment which the Company anticipates will be placed into service by the end of 2019.

Depreciation expense was $508 and $12 during the three months ended June 30, 2019 and 2018, respectively and $979 and $20 during the six months ended June 30, 2019 and 2018, 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 up to 3 years.

On July 24, 2015, Kiniksa US entered into an operating lease in Wellesley Hills, Massachusetts for office space that comprised the former headquarters for Kiniksa US. In March 2016, effective August 1, 2016, Kiniksa US entered into an expansion and extension on its lease, which expanded its leased space to a total of 10,800 square feet. On March 31, 2017, Kiniksa US renewed this lease and extended the lease term to August 2018. Monthly lease payments, inclusive of base rent and ancillary charges, were $27.

On March 13, 2018, Kiniksa US entered into an operating lease in Lexington, Massachusetts for office and laboratory space that comprises the new headquarters for Kiniksa US 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 will be occupied in phases through December 2019. The lease expires on July 31, 2021. Monthly lease

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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 increased from $73 to $101 as of January 1, 2019 and from $101 to $110 as of February 1, 2019. Base rent will increase up to $138 by the earlier of occupation of certain additional expansion space or in December 2019.

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

The components of lease cost consisted of operating lease costs and variable lease costs were $361 and $87 for the three months ended June 30, 2019, respectively and $762 and $109 for the six months ended June 30, 2019, respectively. As of June 30, 2019, the weighted-average lease term was 1.98 years and the discount rate was 7.16%.

Maturities of operating leases liabilities were as follows:

As of June 30,	
2019	$ 767
2020	1,821
2021	972
2022 and thereafter	—
Total future minimum lease payments	$ 3,560
Less imputed interest	(245)
Present value of lease liabilities	$ 3,315

Prior to the adoption of the new lease accounting standard, undiscounted future minimum rents payable as of December 31, 2018 under non-cancelable leases with the initial term exceeding one year were as follows:

As of December 31,	
2019	$ 1,394
2020	1,821
2021	972
2022 and thereafter	—
Total future minimum lease payments	$ 4,187

6. Accrued Expenses

Accrued expenses consisted of the following:

	June 30, 2019	December 31, 2018
Accrued employee compensation and benefits	$ 3,983	$ 5,678
Accrued research and development expenses	13,709	9,656
Accrued legal and professional fees	1,128	994
Other	359	90
	$ 19,179	$ 16,418

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7. Convertible Preferred Shares

As of December 31, 2017, the Company's bye-laws, as amended and restated, designated 22,885,492 authorized shares to be issued as convertible preferred shares with a par value of $0.000273235 per share, of which 17,128,120 shares were further designated as Series A convertible preferred shares (the "Series A preferred shares") and 5,757,372 shares were further designated as Series B convertible preferred shares (the "Series B preferred shares"). In February 2018, the Company's bye-laws were further amended and restated to, among other things, effect an increase in the number of authorized convertible preferred shares with a par value of $0.000273235 per share to 35,670,093 shares, of which 12,784,601 shares were further designated as Series C convertible preferred shares (the "Series C preferred shares"). The holders of convertible preferred shares had liquidation rights in the event of a deemed liquidation that, in certain situations, was not solely within the control of the Company. Therefore, the Series A, Series B and Series C convertible preferred shares (collectively, the "Preferred Shares") were classified outside of shareholders' equity.

In October 2015, the Company issued and sold 8,028,809 Series A preferred shares at a price of $4.6707 per share (the "Series A Original Issue Price") for proceeds of $37,398, net of issuance costs of $102.

In September 2016, the Company issued and sold an additional 9,099,311 Series A preferred shares at a price of $4.6707 per share for proceeds of $42,499, net of issuance costs of $1.

In March 2017, the Company issued and sold 5,757,372 Series B preferred shares at a price of $6.9475 per share (the "Series B Original Issue Price") for proceeds of $39,873, net of issuance costs of $127.

In February 2018, the Company issued and sold 12,784,601 Series C preferred shares at a price of $15.6438 per share (the "Series C Original Issue Price") for proceeds of $190,822, net of issuance costs of $9,178.

In May 2018, upon the completion of the IPO (which qualified as a "Qualified IPO" under the Company's bye-laws, as amended and restated), all of the outstanding Preferred Shares were converted into 5,546,019 Class A common shares, 1,070,502 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares in accordance with the Company's bye-laws, as amended and restated. In connection with the completion of the IPO in May 2018, the Company adopted the Amended & Restated Bye-Laws to, among other things, authorize the issuance of undesignated preferred shares. As of June 30, 2019, no preferred shares were designated or issued.

Prior to the conversion to common shares, the holders of the Preferred Shares had the following rights and preferences:

Voting

The holders of Preferred Shares were entitled to vote, together with the holders of common shares, on all matters submitted to shareholders for a vote. The holders of Series A preferred shares were entitled to the number of votes per Series A preferred share equal to the number of whole Class B common shares into which the Series A preferred shares were convertible at the time of such vote (which was ten votes for each Class B common share). The holders of Series B preferred shares were entitled to the number of votes per Series B preferred share equal to the number of whole Class A common shares into which the Series B preferred shares were convertible at the time of such vote (which was one vote for each Class A common share). The holders of Series C preferred shares were entitled to the number of votes per Series C preferred share equal to the number of whole Class A common shares into which the Series C preferred shares were convertible at the time of such vote (which was one vote for each Class A common share). Except as provided by law or by the other provisions of the Company's bye-laws, holders of Preferred Shares voted together with the holders of common shares as a single class.

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The holders of Preferred Shares, voting together as a single class, were entitled to elect two directors of the Company. The holders of Preferred Shares, voting together with the holders of common shares as a single class, were entitled to elect the remaining directors of the Company, except for the one director that the holders of Class A common shares and Class B common shares, voting together as a single class were entitled to elect.

Conversion

Each Series A preferred share was convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as permitted by Bermuda law, into such number of fully paid and non-assessable Class B common shares determined by dividing the Series A Original Issue Price by the Series A Conversion Price (as defined below) in effect at the time of conversion. Each Series B preferred share was convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as permitted by Bermuda law, into such number of fully paid and non-assessable Class A common shares determined by dividing the Series B Original Issue Price by the Series B Conversion Price (as defined below) in effect at the time of conversion. Each Series C preferred share was convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as permitted by Bermuda law, into such number of fully paid and non-assessable Class A common shares determined by dividing the Series C Original Issue Price by the Series C Conversion Price (as defined below) in effect at the time of conversion.

At the time of the IPO: the Series A Original Issue Price and Series A Conversion Price were equal to $4.6707; the Series B Original Issue Price and Series B Conversion Price were equal to $6.9475; and the Series C Original Issue Price and Series C Conversion Price were equal to $15.6438. Accordingly, each Series A preferred share was convertible into one Class B common share, each Series B preferred share was convertible into one Class A common share and each Series C preferred share was convertible into one Class A common share.

Further, upon either (i) the closing of the sale of Class A common shares or Class B common shares to the public at a price of at least $15.6438 per share (subject to appropriate adjustment in the event of any share dividend, share split, combination or other similar recapitalization with respect to the applicable class of common shares) in an initial public offering resulting in at least $100,000 of gross proceeds to the Company (a “Qualified IPO”) or (ii) the date and time, or the occurrence of an event, specified by vote or written consent of the holders of a majority of the outstanding Preferred Shares, voting together as a single class on an as if converted to Class A common shares basis, all outstanding Series A preferred shares would automatically be converted, in such manner as permitted pursuant to Bermuda law, into Class B common shares at the then effective conversion rate, and all outstanding Series B and Series C preferred shares would automatically be converted, in such manner as permitted pursuant to Bermuda law, into Class A common shares at the then effective conversion rate. Notwithstanding the foregoing, in the event of a mandatory conversion of preferred shares as a result of a Qualified IPO, (a) holders of Series A preferred shares could have elected to receive Class B1 common shares in lieu of Class B common shares and (b) holders of Series B and Series C preferred shares could have elected to receive Class A1 common shares in lieu of Class A common shares.

Dividends

The holders of the Preferred Shares were entitled to receive noncumulative dividends when and if declared by the Company’s board of directors. The Company was not permitted to declare, pay or set aside any dividends on any other class or series of shares of the Company, other than dividends on common shares payable in common shares, unless the holders of the Preferred Share first received, or simultaneously received, a dividend on each outstanding Preferred Share equal to (i) in the case of a dividend on any class of common shares or any class or series was convertible into common shares, that dividend per Preferred Share as would have equaled the product of (a) the dividend payable on each share of such class or series determined, if applicable, as if all shares of such class or series

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had been converted into common shares and (b) the number of common shares issuable upon conversion of a share of the applicable series of Preferred Shares, or (ii) in the case of a dividend on any class or series that was not convertible into common shares, at a rate per Preferred Share determined by (a) dividing the amount of the dividend payable on each share of such class or series of shares by the original issue price of such class or series (subject to appropriate adjustment in the event of any bonus share, share dividend, share split, combination of or other similar recapitalization with respect to such class or series) and (b) multiplying such fraction by an amount equal to the applicable Series A, Series B or Series C Original Issue Price. No cash dividends were declared or paid on the Preferred Shares.

Liquidation Preference

In the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company or deemed liquidation event (as defined below), the holders of Preferred Shares then outstanding were entitled to be paid out of the assets of the Company available for distribution to its shareholders, on a *pari passu* basis, before any payment was made to the holders of common shares by reason of their ownership thereof, an amount per share equal to the greater of (i) one times the applicable Original Issue Price, plus any dividends declared but unpaid thereon, and (ii) such amount per share as would have been payable had all Preferred Shares been converted into common shares immediately prior to such liquidation, dissolution, winding up or deemed liquidation event. Thereafter, the remaining assets of the Company available for distribution to its shareholders would have been distributed among the holders of common shares, pro rata based on the number of shares held by each such holder.

If upon any such liquidation, dissolution or winding up of the Company or deemed liquidation event, the assets of the Company available for distribution to its shareholders were insufficient to pay the holders of Preferred Shares the full amount to which they were entitled, the holders of Preferred Shares would have shared ratably in any distribution of the assets available for distribution in proportion to the respective amounts which would otherwise have been payable in respect of the shares held by such holders of Preferred Shares upon such distribution if all amounts payable on or with respect to such shares were paid in full.

Unless a majority of the holders of the then outstanding Preferred Shares elected otherwise, a deemed liquidation event included a merger or consolidation (other than one in which shareholders of the Company owned a majority by voting power of the outstanding shares of the surviving or acquiring company or corporation) or a sale, lease, transfer, exclusive license or other disposition of all or substantially all of the assets of the Company.

Redemption

The Company's bye-laws, as amended and restated, did not provide redemption rights to the holders of Preferred Shares.

8. Common Shares

As of December 31, 2017, the Company's bye-laws, as amended and restated, authorized the Company to issue 43,918,239 total shares with a par value of $0.000273235, of which 5,507,938 and 3,568,353 shares were designated as Class A and Class B common shares, respectively. In February 2018, the Company's bye-laws were further amended and restated to, among other things, effect an increase in the number of authorized common shares to 44,746,463 shares, of which 5,507,938 shares were designated as Class A common shares and 3,568,353 shares were designated as Class B common shares. The remaining 11,956,456 shares that were not designated as common shares or Preferred Shares as of December 31, 2017 could have been designated to any class at any time in the future by the Company's board of directors. No Class A1 common shares or Class B1 common shares were designated as of December 31, 2017.

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On May 23, 2018, the Company's registration statement on Form S-1 relating to the IPO was declared effective by the SEC. On May 29, 2018, the Company completed the IPO of 8,477,777 Class A common shares at a public offering price of $18.00 per share for gross proceeds of $152,600. In addition, on June 22, 2018, the Company completed the sale of 1,006,425 Class A common shares to the underwriters of the IPO following the exercise in part of their over-allotment option to purchase additional shares at a public offering price of $18.00 per share for gross proceeds of $18,116. The aggregate net proceeds to the Company from the IPO, inclusive of the over-allotment option exercise, was $155,536 after deducting underwriting discounts and commissions and other offering costs.

In May 2018, upon completion of the IPO (which qualified as a "Qualified IPO" under the Company's bye-laws, as amended and restated), all outstanding Preferred Shares were converted into 5,546,019 Class A common shares, 1,070,502 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares in accordance with the Company's bye-laws, as amended and restated. In connection with the completion of the IPO in May 2018, the Company increased the authorized capital of the Company to $54,647 consisting of 200,000,000 shares of $0.000273235 par value per share and, among other things, amended the description of different classes of shares under the Amended & Restated Bye-Laws.

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 $82,985 after deducting underwriting discounts and commissions and other offering costs.

The rights of the holders of the Company's Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares are identical, except with respect to voting and conversion, as described below. As of December 31, 2017, the voting, dividend and liquidation rights of the holders of the Company's common shares were subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares as set forth above. In May 2018, following the conversion of the Preferred Shares into common shares, the voting, dividend and liquidation rights of the holders of the Company's common shares were then subject to and qualified by the rights, powers and preferences of the holders of the preferred shares, if any. As of June 30, 2019, 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. Holders of Class A1 common shares and Class B1 common shares have no voting rights. As of December 31, 2017, the holders of Class A and Class B common shares, voting together as a single class, were entitled to elect one director of the Company. The holders of Class A and Class B common shares, voting together as a single class with the holders of the Preferred Shares, were entitled to elect the remaining directors of the Company, except for the two directors of the Company that the holders of the Preferred Shares, voting together as a single class, were entitled to elect. In May 2018, following the conversion of the Preferred Shares into common shares, 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.

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Dividends

Common shareholders are entitled to receive dividends, as may be declared by the Company's board of directors. As of December 31, 2017, any such dividends would have been subject to the preferential dividend rights of the holders of the Preferred Shares. In May 2018, following the conversion of the Preferred Shares into common shares, any such dividends would be subject to the rights, powers and preferences of any preferred shares that may be issued by the Company. As of June 30, 2019, no preferred shares were designated or issued. Through June 30, 2019, 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. 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. There are no conversion rights associated with the Class A common shares.

9. Share-Based Compensation

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 options, nonqualified 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").

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.

Options

Stock option activity under the Plans is summarized as follows:

	Number of Shares	Weighted Average Fair Value
Outstanding as of December 31, 2018	5,960,939	$ 6.98
Granted	1,808,688	$ 12.35
Exercised	(91,640)	$ 2.60
Forfeited	(302,731)	$ 12.56
Outstanding as of June 30, 2019	7,375,256	$ 8.12

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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 six months ended June 30, 2019 and 2018 were as follows, presented on a weighted-average basis:

	Three Months Ended June 30,		Six Months Ended June 30,	
	2019	2018	2019	2018
Risk-free interest rate	2.16 %	2.87 %	2.48 %	2.72 %
Expected term (in years)	5.97	5.95	6.18	6.51
Expected volatility	78.88 %	74.83 %	78.35 %	75.22 %
Expected dividend yield	— %	— %	— %	— %

Share-Based Compensation

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

	Three Months Ended June 30,		Six Months Ended June 30,	
	2019	2018	2019	2018
Research and development expenses	$ 1,504	$ 363	$ 2,691	$ 565
General and administrative expenses	1,960	696	3,666	1,052
	$ 3,464	$ 1,059	$ 6,357	$ 1,617

Restricted Shares

Under terms of the Class A and Class B restricted share agreements covering the Class A and Class B common shares, restricted common shares are subject to a vesting schedule. The restricted shares vest over a four-year period during which time the Company has the right to repurchase up to all unvested shares at the amount paid if the relationship between the recipient and the Company ceases. Subject to the continued employment (or other engagement of the recipient by the Company as described in the restricted share agreements), all of the restricted common shares become fully vested within four years of the date of issuance.

The following table summarizes restricted share activity for the six months ended June 30, 2019:

	Class A		Class B	
	Number of Shares	Weighted Average Grant Date Fair Value	Number of Shares	Weighted Average Grant Date Fair Value
Unvested restricted shares outstanding as of December 31, 2018	133,812	$0.000273235	743,407	$0.000273235
Granted	—	—	—	—
Vested	(104,076)	$0.000273235	(594,726)	$0.000273235
Unvested restricted shares outstanding as of June 30, 2019	29,736	$0.000273235	148,681	$0.000273235

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10. License and Acquisition Agreements

Biogen Asset Purchase Agreement

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716 and other antibodies covered by certain patent rights, including patents and other intellectual property rights, clinical data, know-how, and clinical drug supply. In addition, Biogen granted to the Company a non-exclusive, sublicensable, worldwide license to certain background patent rights related to the KPL-716 program. The Company is obligated to use commercially reasonable efforts to develop and commercialize such acquired products.

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

Under the Biogen Agreement, the Company is obligated to make milestone payments to Biogen of up to $179,000 upon the achievement of specified clinical and regulatory milestones in multiple indications in various territories, including milestone payments of $4,000 and $10,000 paid during the year ended December 31, 2017 and the six months ended June 30, 2019, respectively, each payment was associated with the achievement of a specified clinical milestone event. Additionally, the Company could be obligated to make up to an aggregate of up to $150,000 of payments upon the achievement of specified annual net sales milestones and to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

The Company also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to the KPL-716 program. Under these retained contracts, the Company paid a one-time upfront sublicense fee of $150 and is obligated to pay insignificant annual maintenance fees as well as clinical and regulatory milestone payments of up to an aggregate of $1,575.

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 six months ended June 30, 2019, the Company recorded research and development expense of $164 and $10,166, respectively, primarily related to milestone payment and other payments associated with the achievement of a specified clinical milestone event due under the Biogen Agreement. During the three and six months ended June 30, 2018, the Company recorded research and development expense of $19 and $30 in connection with annual maintenance fees for the retained contracts 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 has been granted an exclusive, sublicensable, worldwide license under certain intellectual property rights controlled by Novo Nordisk to make, use, develop and commercialize KPL-045 for all indications. The Company is obligated to use commercially reasonable efforts to develop and commercialize such licensed products.

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

Under the Novo Nordisk Agreement, the Company was also required to make a payment of $150 upon completion of the technology transfer by Novo Nordisk. The technology was transferred during the six months ended June 30, 2018 and, as a result, this payment was made and is recorded in the Company's consolidated statement of operations for the six months ended June 30, 2018. In addition, the Company is obligated to make milestone payments upon the achievement of specified clinical, regulatory and initial sales milestones and upon the achievement of annual net sales thresholds, including a payment of $1,000 upon the earlier to occur of a specified regulatory milestone and January 2020, unless the Novo Nordisk Agreement is earlier terminated by either party. As of June 30, 2019 and December 31, 2018, the Company determined that the payment related to the milestone was not probable and, therefore, no amount was recorded in the Company's consolidated statement of operations and comprehensive loss during the three and six months ended June 30, 2019 and 2018. The Company has also agreed to pay royalties on annual net sales of products licensed under the agreement.

Under the Novo Nordisk Agreement, the Company is solely responsible for all development, regulatory and commercial activities and costs. The Company is also responsible for costs related to filing, prosecuting and maintaining the licensed patent rights.

The Novo Nordisk Agreement will terminate upon expiration of the last-to-expire royalty term for any licensed product in the territories, as defined in the agreement. Either party may terminate the agreement upon the other party's insolvency or bankruptcy or for uncured material breach of the agreement by the other party. Novo Nordisk has the right to terminate the agreement if the Company challenges any of the licensed patent rights. The Company may also terminate the agreement for any reason upon prior written notice to Novo Nordisk.

The Company did not incur any research and development expense directly related to milestone payments due under the Novo Nordisk Agreement during the three and six months ended June 30, 2019 and the three months ended June 30, 2018. During the six months ended June 30, 2018 the Company recorded research and development expense of $150, in connection with the completion of technology transfer payment 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.

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

In January 2019, the Company exercised the call option and in March 2019, the Company acquired all of the issued and outstanding equity securities of Primatope (the "Primatope Acquisition") in exchange for $18,000 comprised of upfront consideration of $10,000 at closing and milestone payments of $5,000,which had been achieved as of the closing date, and a milestone of $3,000, which was achieved 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 amounts) in accordance with the Primatope Agreement. At the closing of the Primatope Acquisition, Primatope became a wholly owned subsidiary of the Company and the acquisition was accounted for as an asset acquisition as it did not meet the definition of a business. The Company recorded the upfront payment and milestone payments as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

During the three months ended June 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 six months ended June 30, 2019, the Company recorded research and development expense of $18,000 related to the Primatope Acquisition. During the three and six months ended June 30, 2018, the Company recorded research and development expense of $250, related to the extension of the option period under the Primatope Agreement.

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 six months ended June 30, 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

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comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

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

Under the Regeneron Agreement, the Company is solely responsible for all development and commercialization activities and costs in its 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 six months ended June 30, 2019, the Company recorded research and development expense of $3,127 and $3,642, respectively, and during the three and six months ended June 30, 2018, the Company recorded research and development expense of $1,201 and $1,577, respectively, related to the purchase of drug materials under this agreement. As of June 30, 2019 and December 31, 2018, the Company has non-cancelable purchase commitments under the clinical supply agreement (see Note 12).

The Regeneron Agreement will expire when the Company is no longer developing or commercializing any licensed product under the Regeneron Agreement. Either party may terminate the agreement upon the other party's insolvency or bankruptcy or for material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches). Regeneron has the right to terminate the agreement if the Company suspends its development or commercialization activities for a consecutive 12-month period or does not grant a sublicense to a third-party to perform such activities, or if the Company challenges any of the licensed patent rights. The Company may terminate the agreement at any time that is 18 months after the effective date of the agreement with 180 days' written notice or with one years' 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 month's written notice.

During the three and six months ended June 30, 2019 and June 30, 2018, the Company recorded research and development expense of $3,127, $3,642, $1,201 and $1,577, respectively, related to the purchase of drug materials under the clinical supply agreement with Regeneron. During the three and six months ended June 30, 2019 and 2018, the Company did not record research and development expense in connection with milestone payments due under the Regeneron Agreement.

MedImmune License Agreement

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

In exchange for these rights, the Company made an upfront payment of $8,000. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and

comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use. In addition, the Company is obligated to make clinical, regulatory and initial sales milestone payments of up to $72,500 in aggregate for the first two indications, including, a $5,000 pass-through payment due upon the achievement of a specified clinical milestone event which was met in the fourth quarter of 2018. Also included is a milestone payment of $10,000 due upon the earlier to occur of a specified regulatory milestone and December 31, 2018, unless the MedImmune Agreement is earlier terminated by either party. As of December 31, 2018 and 2017, the Company determined that the payment related to this milestone was probable and, therefore, recognized research and development expense and an accrued milestone of $10,000 during the year ended December 31, 2017. During the six months ended June 30, 2019, the Company made both the $5,000 and $10,000 milestone payments in accordance with the MedImmune Agreement. In addition, the Company is obligated to make clinical and regulatory milestone payments of up to $15,000 in the aggregate for each subsequent indication. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

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

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

During the three and six months ended June 30, 2019 and 2018, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

11. Net Loss per Share

Net Loss per Share Attributable to Common Shareholders

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

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

	Three Months Ended June 30,		Six Months Ended June 30,	
	2019	2018	2019	2018
Numerator:				
Net loss attributable to common shareholders	$ (37,191)	$ (20,259)	$ (103,012)	$ (36,241)
Denominator:				
Weighted average common shares outstanding—basic and diluted	54,475,476	18,328,402	53,225,710	10,492,474
Net loss per share attributable to common shareholders—basic and diluted	$ (0.68)	$ (1.11)	$ (1.94)	$ (3.45)

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

The Company's potentially dilutive securities, which include options and unvested restricted shares have been excluded from the computation of diluted net loss per share attributable to common shareholders as the effect would be to reduce the net loss per share. 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 June 30,	
	2019	2018
Options to purchase common shares	7,375,256	4,805,037
Unvested restricted shares	178,417	1,412,472
	7,553,673	6,217,509

12. Commitments and Contingencies

License Agreements

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

Manufacturing Commitments

The Company entered into agreements with several contract manufacturing organizations to provide preclinical and clinical trial materials. As of June 30, 2019 and December 31, 2018, the Company had committed to minimum payments under these agreements totaling $7,021 and $12,012, respectively.

Indemnification Agreements

In the ordinary course of business, the Company may provide indemnification of varying scope and terms to vendors, lessors, business partners and other parties with respect to certain matters including, but not limited to, losses arising out of breach of such agreements or from intellectual property infringement claims made by third parties. In addition, the Company has entered into indemnification agreements with members of its board of directors and officers

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that will require the Company, among other things, to indemnify them against certain liabilities that may arise by reason of their status or service as directors or officers. The maximum potential amount of future payments the Company could be required to make under these indemnification agreements is, in many cases, unlimited. To date, the Company has not incurred any material costs as a result of such indemnifications. The Company does not believe that the outcome of any claims under indemnification arrangements will have a material effect on its financial position, results of operations or cash flows, and it did not accrue any liabilities related to such obligations in its consolidated financial statements as of June 30, 2019 and December 31, 2018.

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 appearing elsewhere in this Quarterly Report and our audited consolidated financial statements and related notes for the year ended December 31, 2018 included in our Annual Report on Form 10-K, or the 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 the SEC, our actual results could differ materially from the results, performance or achievements expressed in or implied by these forward-looking statements.

Overview

We are a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have a pipeline of product candidates across various stages of development, focused on autoinflammatory and autoimmune conditions. We believe that our product candidates are grounded in strong biologic rationales or validated mechanisms of action and have the potential to address multiple indications.

Our product candidates include rilonacept, mavrilimumab, KPL-716, KPL-404 and KPL-045.

Our lead candidate is rilonacept, 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. We are enrolling a global, double-blind, placebo-controlled, randomized-withdrawal design, pivotal Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis, named RHAPSODY. We expect top-line results from this trial in the second half of 2020. We also completed an open-label Phase 2 proof-of-concept clinical trial in subjects with both symptomatic recurrent pericarditis as well as other patient subsets within pericarditis, including asymptomatic steroid-dependent subjects with recurrent pericarditis and subjects with post-pericardiotomy syndrome. We expect final data from this trial in the second half of 2019.

Mavrilimumab is a monoclonal antibody that antagonizes granulocyte-macrophage colony stimulating factor. We are evaluating mavrilimumab for the potential treatment of giant cell arteritis, or GCA, a chronic inflammatory disease of the medium-large blood vessels with an estimated U.S. prevalence of approximately 75,000 to 150,000 patients. We are enrolling a randomized, double-blind, placebo-controlled, global Phase 2 proof-of-concept trial for the study of mavrilimumab in GCA. We expect top-line data from this trial in the second half of 2020.

KPL-716 is a monoclonal antibody that simultaneously inhibits the signaling of the cytokines interleukin 31, or IL-31, and oncostatin M, or OSM, by targeting their common receptor subunit, oncostatin M receptor beta, or OSMRβ. We are enrolling a randomized, double-blind, placebo-controlled, Phase 2a clinical trial of KPL-716 in subjects with prurigo nodularis, a chronic inflammatory skin condition with an estimated U.S. prevalence of approximately 300,000 patients. We expect top-line data from this trial in the first half of 2020. We are also enrolling an exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus. This randomized, double-blind, placebo-controlled trial is designed to identify chronic pruritic conditions where signaling through OSMRβ may be playing a role and to investigate the efficacy. safety and tolerability of KPL-716 in reducing the moderate-to-severe pruritus experienced by these subjects. We expect top-line data from this study on a cohort-by-cohort basis throughout 2020. Our Phase 1a/1b clinical trial of KPL-716 in healthy volunteers and subjects with moderate to severe atopic dermatitis experiencing moderate to severe pruritus included a single-dose Phase 1b cohort in subjects experiencing moderate to severe atopic dermatitis. We completed enrollment in a 12 week, repeated single dose cohort of the Phase 1b clinical trial, which was

designed to provide safety and exploratory data on disease response markers. We announced interim data from this cohort in August 2019.

KPL-404 is a monoclonal antibody inhibitor of the CD40 co-stimulatory molecule. We are continuing our preclinical activities with KPL-404 in T-cell dependent, B-cell mediated diseases, and expect to file an investigational new drug application, or IND, with the U.S. Food and Drug Administration, or FDA, for this program in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020.

KPL-045, is a monoclonal antibody inhibitor of the CD30L co-stimulatory molecule. We are evaluating the progression of KPL-045 based on preclinical data from the program in the context of our portfolio.

Since our inception in 2015, we have devoted substantially all of our efforts and financial resources to organizing and staffing our company, business planning, raising capital, acquiring, in-licensing or discovering product candidates and securing related intellectual property rights and conducting research and development activities for our programs. We do not have any products approved for sale and have not generated any revenue from product sales. We may never be able to develop or commercialize a marketable product. We have not yet successfully completed any Phase 3 or other pivotal clinical trials, obtained any regulatory approvals, manufactured a commercial-scale drug, or conducted sales and marketing activities.

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 $103.0 million and $36.2 million for the six months ended June 30, 2019 and 2018, respectively. As of June 30, 2019, we had an accumulated deficit of $297.3 million. We expect to continue to incur significant operating losses for at least the next several years as we advance our product candidates through all stages of development and clinical trials and, ultimately, seek regulatory approval. In addition, if we obtain marketing approval for any of our product candidates, we expect to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates. We expect to continue to incur additional costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses.

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

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

As of June 30, 2019, we had cash, cash equivalents and short-term investments of $287.4 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 product sales and do not expect to generate any revenue from the sale of products in the foreseeable future. If our development efforts for our product candidates are successful and result in regulatory approval, we may generate revenue in the future from product sales.

Operating Expenses

Research and Development Expenses

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

- expenses incurred to conduct the necessary 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;
- other costs related to acquiring and manufacturing preclinical studies 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 as well as for managing our preclinical development, process development, manufacturing and clinical development activities.

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

	Three Months Ended June 30,		Six Months Ended June 30,	
	2019	2018	2019	2018
	(in thousands)		(in thousands)	
Rilonacept	$ 8,932	$ 2,903	$ 14,296	$ 4,126
Mavrilimumab	3,136	2,465	8,710	2,743
KPL-716 [1]	6,577	6,712	23,024	13,928
KPL-404 [2]	1,177	671	20,603	1,046
KPL-045	607	890	2,505	1,327
Unallocated research and development expenses	10,419	3,559	20,963	6,661
Total research and development expenses	$30,848	$17,200	$ 90,101	$ 29,831

(1) The amount for the six months ended June 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 six months ended June 30, 2019, includes expense of $18.0 million related to our acquisition of the issued and outstanding equity securities of Primatope, comprised of upfront consideration of $10.0 million at closing, and milestone payments of $5.0 million, which had been achieved as of the closing date, and $3.0 million, which was achieved 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 amounts) in accordance with the Primatope Agreement.

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

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

- the scope, progress, outcome and costs of our preclinical development activities, clinical trials and other research and development activities;
- establishing an appropriate safety and efficacy profile with IND-enabling and clinical studies;
- successful patient enrollment in and the initiation and completion of clinical trials;
- the timing, receipt and terms of any marketing approvals from applicable regulatory authorities including the FDA;
- the extent of any required post-marketing approval commitments to applicable regulatory authorities;
- establishing clinical and commercial manufacturing capabilities or making arrangements with third-party manufacturers in order to ensure that we or our third-party manufacturers are able to make product successfully;

- development and timely delivery of clinical-grade and commercial-grade drug formulations that can be used in our clinical trials and for commercial launch;
- obtaining, maintaining, defending and enforcing patent claims and other intellectual property rights;
- significant and changing government regulation;
- launching commercial sales of our product candidates, if and when approved, whether alone or in collaboration with others; and
- maintaining a continued acceptable safety profile of 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 increase our headcount to support our continued research activities and development of our product candidates and prepare for potential commercialization activities. We also anticipate that we will incur increased accounting, audit, legal, regulatory, compliance and director and officer insurance costs as well as investor and public relations expenses associated with being a public company.

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 will be available to us for those losses. Our wholly owned U.S. subsidiary, Kiniksa Pharmaceuticals Corp., or Kiniksa US, is subject to federal and state income taxes in the United States. Our provision for income taxes relates to taxable income generated by Kiniksa US under a cost-plus arrangement that it has with us.

As of December 31, 2018, we had state research and development tax credit carryforwards of approximately $0.1 million available to reduce future tax liabilities, which begin to expire in 2033.

Results of Operations

Comparison of the Three Months Ended June 30, 2019 and 2018

The following table summarizes our results of operations for the three months ended June 30, 2019 and 2018:

	Three Months Ended June 30,		
	2019	2018	Change
	(in thousands)		
Operating expenses:			
Research and development	$ 30,848	$ 17,200	$ 13,648
General and administrative	8,441	4,327	4,114
Total operating expenses	39,289	21,527	17,762
Loss from operations	(39,289)	(21,527)	(17,762)
Interest income	1,724	1,066	658
Loss before benefit for income taxes	(37,565)	(20,461)	(17,104)
Benefit for income taxes	374	202	172
Net loss	$ (37,191)	$ (20,259)	$ (16,932)

Research and Development Expenses

	Three Months Ended June 30,		
	2019	2018	Change
	(in thousands)		
Direct research and development expenses by program:			
Rilonacept	$ 8,932	$ 2,903	$ 6,029
Mavrilimumab	3,136	2,465	671
KPL-716	6,577	6,712	(135)
KPL-404	1,177	671	506
KPL-045	607	890	(283)
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	7,578	2,816	4,762
Other	2,841	743	2,098
Total research and development expenses	$ 30,848	$ 17,200	$ 13,648

Research and development expenses were $30.8 million for the three months ended June 30, 2019, compared to $17.2 million for the three months ended June 30, 2018. The increase of $13.6 million was due to an increase in external fees related to our development programs in connection with our preclinical and clinical research and development, including procurement of clinical drug supply as well as manufacturing and development of clinical drug supply.

The direct costs for our rilonacept program were $8.9 million during the three months ended June 30, 2019, compared to $2.9 million during the three months ended June 30, 2018, or an increase of $6.0 million. During the three months ended June 30, 2019, expenses incurred primarily related to our clinical research and development for our global, pivotal Phase 3 clinical trial in recurrent pericarditis, including purchases of drug materials under our clinical supply agreement with Regeneron, compared to the three months ended June 30, 2018, in which expenses incurred were primarily related to our clinical research and development for our open-label Phase 2 proof-of-concept clinical trial.

The direct costs for our mavrilimumab program were $3.1 million during the three months ended June 30, 2019, compared to $2.5 million during the three months ended June 30, 2018, or an increase of $0.6 million. During the three months ended June 30, 2019, expenses incurred related primarily to our global Phase 2 clinical trial in GCA and manufacturing process development related expenses compared to the three months ended June 30, 2018, in which expenses incurred related primarily to preparation for our global Phase 2 clinical trial in GCA, including manufacturing development costs.

The direct costs for our KPL-716 program were $6.6 million during the three months ended June 30, 2019, compared to $6.7 million during the three months ended June 30, 2018, or a decrease of $0.1 million. During the three months ended June 30, 2019, expenses incurred primarily related to our Phase 2a clinical trial in prurigo nodularis and trial initiation costs for our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus, including manufacturing and development costs for our clinical drug supply compared to the three months ended June 30, 2018, in which expenses incurred related primarily to our Phase 1a/1b clinical trial and our LOTUS-PN observational study, including manufacturing and development costs for our clinical drug supply.

The direct costs for our KPL-404 program were $1.2 million during the three months ended June 30, 2019, compared to $0.7 million during the three months ended June 30, 2018, or an increase of $0.5 million. During the three months ended June 30, 2019, expenses incurred primarily related to preclinical research and development, including manufacturing development costs. During the three months ended June 30, 2018, expenses incurred related to preclinical research and development, including manufacturing development costs as well as expenses related to the extension of the option period under the Primatope Agreement.

The direct costs for our KPL-045 program were $0.6 million during the three months ended June 30, 2019, compared to $0.9 million during the three months ended June 30, 2018, or a decrease of $0.3 million. During the three months ended June 30, 2019, expenses incurred related to preclinical research and development, compared to the three months ended June 30, 2018, in which expenses incurred related to preclinical research and development, including manufacturing development costs.

Unallocated research and development expenses were $10.4 million for the three months ended June 30, 2019 compared to $3.6 million for the three months ended June 30, 2018. The increase of $6.8 million in unallocated research and development expenses was due to an increase of $4.7 million in personnel-related costs, including share-based compensation, and an increase of $2.1 million in other costs, including operating costs for our laboratory as well as research costs related to potential future programs. The increase in personnel-related costs was primarily due to the hiring of additional personnel in our research and development functions, particularly those responsible for coordinating with CMOs on process development and manufacturing of drug supply and coordinating with CROs on the conduct and oversight of our current and planned clinical trials as well as research studies and development programs for our product candidates. Personnel-related costs for the three months ended June 30, 2019 and 2018 included share-based compensation of $1.5 million and $0.4 million, respectively.

General and Administrative Expenses

General and administrative expenses were $8.4 million for the three months ended June 30, 2019 compared to $4.3 million for the three months ended June 30, 2018. The increase of $4.1 million was due to increases of $2.9 million in personnel-related costs and $1.2 million in professional fees and other expenses. The increase in personnel-related costs was due to the hiring of additional personnel in our general and administrative functions, primarily in our corporate departments, including legal, finance and human resources, as we continued to expand our operations to support the organization and our status as a public company. Personnel-related costs for the three months ended June 30, 2019 and 2018 included share-based compensation of $2.0 million and $0.7 million, respectively. Professional fees increased due to legal costs incurred in connection with maintaining and registering worldwide patents and costs associated with our ongoing business operations, as well as higher accounting, recruiting, market research expenses and other costs incurred due to being a public company.

Interest Income

Interest income was $1.7 million for the three months ended June 30, 2019 compared to $1.1 million for the three months ended June 30, 2018. The increase was due to both higher average invested balances and higher interest rates on U.S. Treasury notes.

Benefit for Income Taxes

For the three months ended June 30, 2019 and 2018, we recorded an insignificant benefit for income taxes.

Comparison of the Six Months Ended June 30, 2019 and 2018

The following table summarizes our results of operations for the six months ended June 30, 2019 and 2018:

	Six Months Ended June 30,		
	2019	2018	Change
	(in thousands)		
Operating expenses:			
Research and development	$ 90,101	$ 29,831	$ 60,270
General and administrative	16,835	8,036	8,799
Total operating expenses	106,936	37,867	69,069
Loss from operations	(106,936)	(37,867)	(69,069)
Interest income	3,533	1,371	2,162
Loss before benefit for income taxes	(103,403)	(36,496)	(66,907)
Benefit for income taxes	391	255	136
Net loss	$ (103,012)	$ (36,241)	$ (66,771)

Research and Development Expenses

	Six Months Ended June 30,		
	2019	2018	Change
	(in thousands)		
Direct research and development expenses by program:			
Rilonacept	$ 14,296	$ 4,126	$ 10,170
Mavrilimumab	8,710	2,743	5,967
KPL-716	23,024	13,928	9,096
KPL-404	20,603	1,046	19,557
KPL-045	2,505	1,327	1,178
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	14,514	5,273	9,241
Other	6,449	1,388	5,061
Total research and development expenses	$ 90,101	$ 29,831	$ 60,270

Research and development expenses were $90.1 million for the six months ended June 30, 2019, compared to $29.8 million for the six months ended June 30, 2018. The increase of $60.3 million was primarily due to an increase in external fees related to our development programs, the primary drivers of the increase included $18.0 million of expense related to the acquisition of the issued and outstanding equity securities of Primatope, comprised of upfront consideration of $10.0 million paid at closing and milestone payments of $5.0 million, which had been achieved as of the closing date, and $3.0 million, which was achieved 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 amounts) in accordance with the Primatope Agreement. Also, contributing to the increase was $10.0 million related to an accrued milestone under our asset purchase agreement with Biogen associated with the achievement of a specified clinical milestone event. In addition, we had an increase in external spend for our programs of approximately $18.0 million as well as an increase of $14.3 million in unallocated research and development expenses.

The direct costs for our rilonacept program during the six months ended June 30, 2019 were $14.3 million, compared to $4.1 million during the six months ended June 30, 2018, or an increase of $10.2 million. During the six months ended June 30, 2019, expenses incurred primarily related to our clinical research and development for our global pivotal Phase 3 clinical trial in recurrent pericarditis, including 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 compared to the six months ended June 30, 2018, in which expenses incurred were primarily related to clinical research and development

with our open-label Phase 2 proof-of-concept clinical trial and preparation for our global pivotal Phase 3 clinical trial in recurrent pericarditis.

The direct costs for our mavrilimumab program during the six months ended June 30, 2019 were $8.7 million, compared to $2.7 million during the six months ended June 30, 2018, or an increase of $6.0 million. During the six months ended June 30, 2019, expenses incurred related primarily to our global Phase 2 trial in GCA and manufacturing process development related expenses compared to the six months ended June 30, 2018, in which expenses related primarily to preparation for our global Phase 2 trial in GCA.

The direct costs of our KPL-716 program were $23.0 million during the six months ended June 30, 2019, compared to $13.9 million during the six months ended June 30, 2018, or an increase of $9.1 million. During the six months ended June 30, 2019, expenses incurred related primarily to a milestone payment of $10.0 million under our asset purchase agreement with Biogen associated with the achievement of a specified clinical milestone event as well as expenses incurred for our Phase 2a clinical trial in prurigo nodularis; our exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus; our Phase 1b clinical trial, as well as manufacturing and development costs for our clinical drug supply, compared to the six months ended June 30, 2018, in which expenses incurred related primarily to our Phase 1a/1b clinical trial and our LOTUS-PN observational study, including manufacturing and development costs for our clinical drug supply.

The direct costs for our KPL-404 program during the six months ended June 30, 2019 were $20.6 million, compared to $1.0 million during the six months ended June 30, 2018, or an increase of $19.6 million. During the six months ended June 30, 2019, expenses incurred primarily related to $18.0 million of expense related to the acquisition of the issued and outstanding equity securities of Primatope, comprised of upfront consideration of $10.0 million paid at closing and milestone payments of $5.0 million, which had been achieved as of the closing date, and $3.0 million, which was achieved 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 amounts) in accordance with the Primatope Agreement. During the six months ended June 30, 2018, expenses incurred related to $0.7 million of direct costs related to clinical research and development, including manufacturing development costs as well as $0.3 million related to the extension of the option period under our stock purchase option agreement with Primatope. The Primatope acquisition was accounted for as an asset acquisition as it did not meet the definition of a business. We recorded the upfront payment, milestone payments and the accrued milestone 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.

The direct costs for our KPL-045 program during the six months ended June 30, 2019 were $2.5 million, compared to $1.3 million during the six months ended June 30, 2018, or an increase of $1.2 million. During the six months ended June 30, 2019, expenses incurred related to preclinical research and development, including manufacturing development costs, compared to the six months ended June 30, 2018, in which expenses incurred related to $1.1 million of direct costs related to preclinical research and development, including manufacturing development costs, as well as $0.2 million related to a technology transfer payment under our license agreement with Novo Nordisk A/S, or NovoNordisk.

Unallocated research and development expenses were $21.0 million for the six months ended June 30, 2019 compared to $6.7 million for the six months ended June 30, 2018. The increase of $14.3 million in unallocated research and development expenses was due to an increase of $9.2 million in personnel-related costs, including share-based compensation, and an increase of $5.1 million in other costs. The increase in personnel-related costs was primarily due to the hiring of additional personnel in our research and development functions, particularly those responsible for coordinating with CMOs on process development and manufacturing of drug supply and coordinating with CROs on the conduct and oversight of our current and planned clinical trials as well as research studies and development programs for our product candidates. Personnel-related costs for the six months ended June 30, 2019 and 2018 included share-based compensation of $2.7 million and $0.6 million, respectively.

General and Administrative Expenses

General and administrative expenses were $16.8 million for the six months ended June 30, 2019 compared to $8.0 million for the six months ended June 30, 2018. The increase of $8.8 million was primarily due to increases of $6.5 million in personnel-related costs and $2.3 million in professional fees and other costs. The increase in personnel-related costs was due to the hiring of additional personnel in our general and administrative functions, primarily in our corporate, finance and human resources departments, as we continued to expand our operations to support the organization, including our status as a public company. Personnel-related costs for the six months ended June 30, 2019 and 2018 included share-based compensation of $3.7 million and $1.1 million, respectively. Professional fees increased due to legal costs incurred in connection with maintaining and registering worldwide patents and costs associated with our ongoing business operations, as well as higher accounting, recruiting, market research expenses and other costs incurred due to being a public company.

Interest Income

Interest income was $3.5 million for the six months ended June 30, 2019 compared to $1.4 million for the six months ended June 30, 2018. The increase was due to both higher average invested balances and higher interest rates on U.S. Treasury notes.

Benefit for Income Taxes

We recorded an insignificant benefit for income taxes for the six months ended June 30, 2019 and 2018.

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. Prior to the completion of the IPO in May 2018, we funded our operations primarily with proceeds from the sale of preferred shares, from which we had received net proceeds of $310.6 million.

On May 29, 2018, we completed the IPO of 8,477,777 Class A common shares at a public offering price of $18.00 per share for gross proceeds of $152.6 million. In addition, on June 22, 2018, we completed the sale of 1,006,425 Class A common shares to the underwriters of the IPO following the exercise in part of their over-allotment option to purchase additional shares at a public offering price of $18.00 per share for gross proceeds of $18.1 million. The aggregate net proceeds to us from the IPO, inclusive of the over-allotment option exercise, was $155.5 million after deducting underwriting discounts and commissions and other offering costs.

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

As of June 30, 2019, we had cash, cash equivalents and short-term investments of $287.4 million.

Cash Flows

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

	Six Months Ended June 30,	
	2019	2018
	(in thousands)	
Net cash used in operating activities	$ (104,932)	$ (32,889)
Net cash provided by (used in) investing activities	32,085	(73,987)
Net cash provided by financing activities	83,681	347,207
Net increase in cash and cash equivalents and restricted cash	$ 10,834	$ 240,331

Operating Activities

During the six months ended June 30, 2019, operating activities used $104.9 million of cash, primarily resulting from our net loss of $103.0 million, partially offset by non-cash charges of $12.7 million and net cash used by changes in our operating assets and liabilities of $14.6 million. Net cash used by changes in our operating assets and liabilities for the six months ended June 30, 2019 consisted of a $15.0 million decrease in accrued milestones, a $1.6 million decrease in accounts payable, a $0.6 million decrease in operating lease liabilities, a $1.0 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. The decrease in accounts payable was primarily due to the timing of vendor invoicing and payments. The increase in prepaid expenses and other current assets was due to interest receivable and prepaid expenses to CMOs related to manufacturing development and CROs related to our clinical trials. The decrease in operating lease liabilities is due to monthly payments for our right of use assets.

During the six months ended June 30, 2018, operating activities used $32.9 million of cash, primarily resulting from our net loss of $36.2 million, partially offset by non-cash charges of $1.4 million and net cash provided by changes in our operating assets and liabilities of $1.9 million. Net cash provided by changes in our operating assets and liabilities for the six months ended June 30, 2018 consisted of a $5.6 million increase in accrued expenses partially offset by a $3.0 million increase in prepaid expenses and other current assets and a $0.7 million decrease in accounts payable. The increase in accrued expenses was due to our increased level of operating activities and the timing of vendor invoicing and payments as well as an increase in accrued employee compensation expense. The increase in prepaid expenses and other current assets was due to increases in prepaid insurance expenses, interest receivable and prepaid expenses to CMOs related to manufacturing development and CROs related to our clinical trials. The decrease in accounts payable was primarily due to the timing of vendor invoicing and payments.

Investing Activities

During the six months ended June 30, 2019 investing activities provided $32.1 million of cash, consisting of $306.4 million from proceeds of maturities of short-term investments, partially offset by $0.8 million of purchases of property and equipment and $273.5 million of purchases of short-term investments.

During the six months ended June 30, 2018, investing activities used $74.0 million of cash, consisting of $0.3 million of purchases of property and equipment and $73.7 million of purchases of short-term investments.

Financing Activities

During the six months ended June 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 commissions and discounts and other offering costs.

During the six months ended June 30, 2018, net cash provided by financing activities was $347.2 million, primarily consisting of proceeds of $159.2 million from our issuance and sale of Class A common shares, net of underwriting commissions and discounts upon completion of our IPO and $190.8 million in net proceeds from our issuance and sale of Series C preferred shares, partially offset by $2.8 million of payments of other offering costs associated with our IPO.

Funding Requirements

We expect our expenses to increase substantially in connection with our ongoing activities, particularly as we advance the clinical trials and preclinical activities of our product candidates. Additionally, we expect to continue to incur additional costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses. Our expenses will also increase as we:

- continue to conduct our current clinical trials and/or initiate our planned clinical trials, for rilonacept, mavrilimumab, KPL-716 and KPL-404, as applicable;
- manufacture, or have manufactured on our behalf, our preclinical and clinical drug material and develop processes for late stage and commercial manufacturing;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- establish a sales, marketing, medical affairs, pricing and reimbursement and distribution infrastructure to commercialize any product candidates for which we may obtain marketing approval and intend to commercialize on our own;
- hire additional clinical, quality control and scientific personnel;
- expand our operational, financial and management systems and increase personnel, including personnel globally to support our clinical development, manufacturing and commercialization efforts and our operations as a public company;
- advance preclinical development of our early-stage programs;
- 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, we expect to incur significant commercialization expenses related to product manufacturing, sales, marketing and distribution, depending on where we choose to commercialize.

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

- the scope, progress, results and costs of researching and developing our product candidates, and conducting 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, including grants from organizations and foundations;
- the revenue, if any, received from commercial sale of our products, should any of our product candidates receive marketing approval;
- the costs of preparing, filing and prosecuting patent applications, maintaining and enforcing our intellectual property rights and defending intellectual property-related claims;
- our ability to establish and maintain collaborations on favorable terms, if at all;
- the extent to which we acquire or in-license other product candidates, 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, governmental funding, collaborations, strategic partnerships or marketing, distribution or licensing arrangements with third parties. To the extent that we raise additional capital through the sale of equity or convertible debt securities, 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 governmental funding, collaborations, strategic partnerships or marketing, distribution or licensing arrangements with third parties, we may have to relinquish valuable rights to our technologies, future revenue streams, research programs or product candidates or grant licenses on terms that may not be favorable to us. If we are unable to raise additional funds when needed, we may be required to delay, reduce or eliminate our product development or future commercialization efforts.

Contractual Obligations and Commitments

During the six months ended June 30, 2019, 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, 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 six months ended June 30, 2019, there were no material changes to our critical accounting policies, other than the adoption of the new leasing standard. Our critical accounting policies are described under the heading "Management's Discussion and Analysis of Financial Condition and Results of Operations— Critical Accounting Policies and Significant Judgments and Estimates" in the Annual Report and the notes to the consolidated financial statements included in Item 1, "Consolidated Unaudited Financial Statements," included in this Quarterly Report. We believe that of our critical accounting policies, the following accounting policies involve the most judgment and complexity:

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

Emerging Growth Company Status

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

Off-Balance Sheet Arrangements

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

Item 3. Quantitative and Qualitative Disclosures About Market Risk.

Interest Rate Risk

We are exposed to market risk related to changes in interest rates. As of June 30, 2019, 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. 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 Chief Executive Officer and Chief 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 Chief Executive Officer and Chief Financial Officer concluded that our disclosure controls and procedures were effective at the reasonable assurance level as of June 30, 2019.

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 June 30, 2019 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 unaudited consolidated financial statements and the related notes and "Management's Discussion and Analysis of Financial Condition and Results of Operations", and our other filings made with the Securities and Exchange Commission, or SEC. The occurrence of any of the events or developments described below could adversely affect our business, financial condition, results of operations and growth prospects. In such an event, the market price of our Class A common shares could decline. Additional risks and uncertainties not presently known to us or that we currently deem immaterial also may impair our business operations.

Risks Related to Our Financial Position and Capital Needs

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

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

We have incurred significant losses related to expenses for research and development and our ongoing operations. Our net losses for the six months ended June 30, 2019 and 2018 were $103.0 million and $36.2 million, respectively. As of June 30, 2019, we had an accumulated deficit of $297.2 million. We expect to continue to incur losses for the foreseeable future, and we anticipate these losses will increase substantially as we:

- continue our research and preclinical and clinical development of our product candidates, including our global, pivotal Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, named RHAPSODY, our global Phase 2 clinical trial with mavrilimumab for the treatment of giant cell arteritis, or GCA, our Phase 2a clinical trial with KPL-716 in prurigo nodularis and our exploratory Phase 2 clinical trial for KPL-716 in diseases characterized by chronic pruritus;
- advance the development of our preclinical programs, including our plans to file an investigational new drug application, or IND, for KPL-404 and our evaluation of KPL-045 based on preclinical data in the context of our portfolio;
- initiate other potential additional preclinical studies and clinical trials for our product candidates;
- increase our manufacturing capabilities or add additional manufacturers or suppliers;

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

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

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

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

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

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

- the results from, and the time and cost necessary for, completing our global, pivotal Phase 3 clinical trial for rilonacept in recurrent pericarditis, RHAPSODY, our global Phase 2 clinical trial for mavrilimumab in GCA, our Phase 2a clinical trial for KPL-716 in prurigo nodularis, our exploratory Phase 2 clinical trial for KPL-716 in diseases characterized by chronic pruritus, as well as our planned filing of an IND for KPL-404 and our evaluation of KPL-045 based on preclinical data in the context of our portfolio;
- the number, size and type of preclinical activities and any additional clinical trials;
- the costs, timing and outcomes of seeking and potentially obtaining approvals from the U.S. Food and Drug Administration, or FDA, or comparable foreign regulatory authorities, including the potential for the FDA or comparable regulatory authorities to require that we conduct more studies than those that we currently expect to conduct and the costs of post-marketing studies or risk evaluation and mitigation strategies, or REMS, that could be required by regulatory authorities;
- the costs and timing of transferring manufacturing technology to third-party manufacturers, producing product candidates to support clinical trials and preparing to manufacture mavrilimumab and KPL-716 on a commercial scale, as well as producing rilonacept in potential new final form configurations;
- the timing and amount of milestone and other payments we must make under our agreements with Regeneron Pharmaceuticals, Inc., or Regeneron, MedImmune, Limited, or MedImmune, Biogen MA Inc., or Biogen, Novo Nordisk A/S, or Novo Nordisk, and the other third parties from whom we have acquired or in-licensed our product candidates or from whom we may in the future acquire or in-license product candidates;
- our ability to successfully commercialize any of our product candidates, including the cost and timing of forming and expanding our sales organization and marketing capabilities;
- the amount of sales revenues from our product candidates, if approved, including the sales price and the availability of coverage and adequate third-party reimbursement;
- competitive and potentially competitive products and technologies and patients' receptivity to our product candidates and the technology underlying them in light of competitive products and technologies;
- the cash requirements of any future acquisitions, developments or discovery of additional product candidates, including any licensing, 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 activities;
- the costs associated with being a public company;

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

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

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

Risks Related to Product Development and Regulatory Approval

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

We do not currently generate any revenue from sales of any products, and we may never be able to develop or commercialize marketable products. We have three product candidates in various stages of clinical development and two at the preclinical development stage. 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 cryopyrin-associated periodic syndrome, or CAPS, in the United States by Regeneron, we are studying rilonacept for the treatment of a different indication called recurrent pericarditis, for which we have completed a Phase 2 clinical trial and which is currently in a global, pivotal Phase 3 clinical trial, RHAPSODY. Mavrilimumab has been through Phase 2 clinical trials conducted by MedImmune for the treatment of rheumatoid arthritis, or RA, but our global Phase 2 clinical trial with mavrilimumab is for the treatment of GCA. Our third clinical-stage product candidate, KPL-716, is being studied in a Phase 2a clinical trial in prurigo nodularis and an exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus. We also have preclinical product candidates that would need to progress through studies to enable an IND prior to clinical development. We are not

permitted to market or promote any of our product candidates before we receive regulatory approval from the FDA or comparable foreign regulatory authorities. Our assumptions about why these product candidates are worthy of future development and potential approval in these, or any, indications are based on either indirect data primarily collected by other companies or our preclinical and clinical trials.

We have not submitted, and we may never submit marketing applications to the FDA or comparable foreign regulatory authorities for our product candidates. We cannot be certain that any of our product candidates will be successful in clinical trials or receive regulatory approval. Further, our product candidates may not receive regulatory approval even if we complete a successful clinical trial. 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, obtaining manufacturing capacity and expertise, success manufacture of clinical supply, building a commercial organization, substantial investment and significant marketing efforts before we will be able to generate any revenue from product sales. The success of our product candidates depends upon several factors, including the following:

- successful completion of preclinical studies, including toxicology studies, biodistribution studies and minimally efficacious dose studies in animals, conducted, where applicable, under the FDA's Good Laboratory Practice, or GLP, regulations;
- submission to and acceptance by the FDA of INDs and of clinical trial applications to foreign governmental authorities for our product candidates to commence planned clinical trials or future clinical trials;
- successful site activation for and 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;
- establishment of arrangements with third-party manufacturers, as applicable, for continued clinical supply and commercial manufacturing;
- successful development of our manufacturing processes and transfer to new third-party facilities to support future development activities and commercialization that are operated by contract manufacturing organizations, or CMOs, in a manner compliant with all regulatory requirements;
- successful manufacture of sufficient supplies of our product candidates within approved specifications for purity and efficacy from our facility and from our CMOs in order to meet clinical or commercial demand, as applicable;
- establishment and maintenance of patent and trade secret protection or regulatory exclusivity for our product candidates;
- successful commercial launch of our product candidates, if and when approved;

- acceptance of our products, if and when approved, by patients, the medical community and third-party payors;
- effective competition with other therapies;
- establishment and maintenance of adequate healthcare coverage and reimbursement;
- enforcement and defense of intellectual property rights and claims;
- continued compliance with any post-marketing requirements imposed by regulatory authorities, including any required post-marketing clinical trials or REMS; and
- maintenance of a continued acceptable safety profile of our product candidates following approval.

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

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

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

We are enrolling a global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, RHAPSODY, a global Phase 2 clinical trial with mavrilimumab for the treatment of GCA, a Phase 2a clinical trial with KPL-716 in prurigo nodularis, and an exploratory Phase 2 clinical trial with KPL-716 in diseases characterized by chronic pruritus. We are also continuing our preclinical activities with KPL-404 prior to initiating clinical trials and are evaluating the progression of KPL-045 based on preclinical data in the context of our portfolio. Commencing our planned clinical trials is subject to acceptance by the FDA of an IND or an IND amendment, acceptance by European regulatory authorities of a Clinical Trial Application, 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 schedule, 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, based on FDA feedback, we will need to demonstrate the effectiveness and safety of mavrilimumab at the 26 weeks of dosing stipulated in our Phase 2 clinical trial in GCA, and eventually demonstrate effectiveness and safety of mavrilimumab beyond 26 weeks as well as evaluate its pharmacokinetic profile and the effectiveness of mavrilimumab at different doses in GCA.

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

A failure of one or more clinical trials can occur at any stage of testing, and our future clinical trials may not be successful. We may experience delays in our ongoing clinical trials and we do not know whether planned clinical trials will begin on time, will be allowed by regulatory authorities, need to be redesigned, 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 successful or timely completion of clinical development 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 in reaching a consensus with regulatory agencies on trial design or implementation;
- delays in establishing the appropriate dosage levels or frequency of dosing or treatment in clinical trials;
- delays in reaching, or failing to reach, agreement on acceptable terms with prospective CROs, and clinical trial sites, the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and clinical trial sites;
- difficulties in obtaining required institutional review board, or IRB, approval at each clinical trial site;
- delays in or failure to obtain regulatory approval to commence a trial, or imposition of a clinical hold by regulatory agencies, after review of an IND 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 to participate in our clinical trials;
- amendments to clinical trial protocols amending 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, other third parties, or us to adhere to clinical trial requirements or to perform their obligations in a timely or compliant manner;
- failure to perform in accordance with the FDA's good clinical practices requirements, or GCPs, or applicable regulatory guidelines in other countries;
- patients not completing participation in a clinical trial or return for post-treatment follow-up;
- clinical trial sites or patients dropping out of a clinical trial;
- participating patients experiencing serious adverse events or undesirable side effects or 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 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; and
- delays in manufacturing, testing, releasing, validating or importing/exporting sufficient stable quantities of our product candidates for use in clinical trials or the inability to do any of the foregoing.

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

Moreover, principal investigators for our clinical trials may serve as scientific advisors or consultants to us from time to time and receive compensation in connection with such services. Under certain circumstances, we may be required to report some of these relationships to the FDA or other regulatory authorities. 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. This could result in a delay in approval, or rejection, of our marketing applications by the FDA or other regulatory authorities, as the case may be, and may ultimately lead to the denial of marketing approval of our product candidates.

If we experience delays in the completion of any clinical trial of our product candidates or any clinical trial of our product candidates is terminated, the commercial prospects of our product candidates may be harmed, and our ability to generate product revenues from our product candidates, if any, will be delayed. Moreover, any delays in completing our clinical trials will increase our costs, slow down the development and approval process of our product candidates and jeopardize our ability to commence product sales and generate revenue, if any. 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.

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

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

We must produce, through third parties, sufficient stable quantities of our product candidates for use in our clinical trials. Any delays in the production of our product candidates may lead to a delay in our clinical trials. If we make manufacturing or formulation changes to our product candidates or change manufacturers or manufacturing processes, we may be unsuccessful in producing the product as compared to the process or manufacturer used in prior clinical trials, and therefore may need to conduct additional trials to bridge our modified product candidates to earlier versions, which could impact the timing of commencing or completing our clinical trials. Moreover, there is no assurance that future clinical trials utilizing a new formulation of a product candidate manufactured by different manufacturers or pursuant to a new process will result in the favorable result, if any, observed in the prior clinical trials of such product candidates. For example, we will need to produce mavrilimumab using different media and feed compared to the processes that were used by MedImmune to develop our existing inventory. Further, we transferred the manufacturing of mavrilimumab to a third party to manufacture mavrilimumab to supply Phase 3 clinical trials and commercialization efforts, if any. This manufacturer may be unsuccessful in producing the product in quantities or quality necessary to support our clinical trials or commercialization efforts, if any, which would delay development of mavrilimumab. In addition, we built manufacturing capabilities to support certain preclinical and early clinical development for our product candidates. We may not be able to produce sufficient quantities of these product candidates or produce them at an acceptable quality, which could delay, prevent or impair our development or commercialization efforts.

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.

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

Identifying and qualifying patients to participate in clinical trials of our product candidates is critical to our success. The timing of our clinical trials depends in part on the speed at which we can recruit patients to participate in

testing our product candidates, and we may experience delays in our clinical trials if we encounter difficulties in enrollment. Patient enrollment depends on many factors, including the size and nature of the patient population, eligibility criteria for the clinical trial, the proximity of patients to clinical sites, the design of the clinical protocol, the availability of competing clinical trials, the availability of new drugs approved for the indication the clinical trial is investigating, the risk that patients enrolled in clinical trials will drop out of the trials before completion of their treatment and clinicians' and patients' perceptions as to the safety and potential advantages of the product candidate being studied in relation to other available therapies. Many of the conditions for which we plan to evaluate our current product candidates in the near future are in small disease populations. Accordingly, there are limited patient pools from which to draw for clinical trials.

In addition to the rarity of these diseases, the eligibility criteria of our clinical trials will further limit the pool of available trial participants, as we will require patients to have specific characteristics that we can measure or to ensure their disease is either severe enough or not too advanced to include them in a trial. Further, we could learn that our clinical trial design increased the difficulty of enrolling patients, which could delay our trials. The process of finding and diagnosing patients may prove costly, especially since the rare diseases we are studying are commonly under diagnosed. If patients are unwilling to participate in our trials for any reason, the timeline for recruiting patients, conducting trials and obtaining regulatory approval of potential products may be delayed, the commercial prospects of our product candidates will be harmed, and our ability to generate product revenue from any of these product candidates could be delayed or prevented. Moreover, failure to obtain and maintain patient consents can also lead to delay or prevent completion of clinical trials of our product candidates.

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

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

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

All of our product candidates modulate the immune system and carry risks associated with immunosuppression, including the risk of serious infections and cancer. Some common side effects of rilonacept include, cold symptoms, nausea, stomach pain, diarrhea, numbness or tingly feeling and injection-site reaction. IL-1 blockade may interfere with immune response to 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 adverse events, or AEs, were gastrointestinal disorders and injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment.

For mavrilimumab, there is a theoretical risk for the development of pulmonary alveolar proteinosis, or PAP. PAP is a rare lung disorder in which surfactant-derived lipoproteins accumulate excessively within pulmonary alveoli due to loss of 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 foreign regulatory agency 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 foreign regulatory authorities could order us to cease further development of, or deny or withdraw approval of, any of our product candidates for any or all targeted indications.

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

- regulatory authorities may withdraw approvals of such product and require us to take 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.

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

We had no involvement with or control over the preclinical and clinical development of any of our product candidates prior to our in-license or acquisition of them. We are dependent on Regeneron, MedImmune, Biogen, Primatope and Novo Nordisk having conducted their research and development in accordance with the applicable protocols and legal, regulatory and scientific standards; having accurately reported the results of all preclinical studies and clinical trials conducted prior to our in-license or acquisition; and having correctly collected, interpreted, and completely transferred the data from these trials or other studies to us. If these activities were not compliant, accurate or correct, the clinical development, regulatory approval, or commercialization of one or more of our product candidates will be adversely affected.

If we cannot replicate positive results from earlier preclinical studies and clinical trials conducted by us or the companies from whom we have licensed or acquired, or may in the future license or acquire, our product candidates in our later clinical trials, we may be unable to successfully develop, obtain regulatory approval for and commercialize our product candidates.

Positive results from our preclinical studies and any positive results we may obtain from our early clinical trials of our product candidates, or from the clinical trials conducted by the companies from whom we in-licensed or acquired or may in the future in-license or acquire our product candidates, may not necessarily be predictive of the results from any required later preclinical studies and clinical trials. Similarly, even if we are able to complete our planned preclinical studies or clinical trials of our product candidates, the positive results from the preclinical studies and clinical trials of our product candidates may not be replicated in our subsequent preclinical studies or clinical trial results. The mechanisms of action of our product candidates may not prove to be safe or effective to treat the diseases that we are studying. Further, the safety and efficacy of our product candidates have not been established for the indications in which we are developing them, and we cannot provide any assurance that their development will be successful. For example, although rilonacept is FDA approved for the treatment of CAPS, and mavrilimumab has been studied in Phase 2 clinical trials for the treatment of RA, their safety and efficacy have not been determined in the indications we are pursuing, recurrent pericarditis or GCA, respectively, and each may fail to receive regulatory approval for those indications.

Many companies in the pharmaceutical and biotechnology industries have suffered significant setbacks in later-stage clinical trials after achieving positive results in early-stage development and we cannot be certain that we will not face similar setbacks. These setbacks have been caused by, among other things, preclinical findings made while clinical trials were underway or safety or efficacy observations made in preclinical studies and clinical trials, including previously unreported AEs. 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 foreign regulatory agencies may significantly change in a manner rendering our clinical data insufficient for approval, which may lead to the FDA or any foreign regulatory agencies delaying, limiting or denying approval of our product candidates.

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

From time to time, we may publish preliminary, interim or "top-line" data from our clinical trials. For example, in March 2019, we released interim data from the open-label Phase 2 proof-of-concept clinical trial of rilonacept in recurrent pericarditis. Preliminary, interim and "top-line" data from clinical trials that we may complete are subject to the risk that one or more of the clinical outcomes may materially change as patient enrollment continues and more patient data become available. Preliminary, interim and "top-line" data also remain subject to audit and verification procedures that may result in the final data being materially different from the data we previously published. As a result, preliminary, interim and "top-line" data should be viewed with caution until the final data are available. Adverse differences between preliminary, interim or "top-line" data and final data could significantly harm our business prospects.

Regulatory approval processes are lengthy, time consuming and inherently unpredictable. If we are not able to obtain, or if there are delays in obtaining, required regulatory approvals for our product candidates 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 design, testing, manufacture, safety, efficacy, recordkeeping, labeling, storage, approval, advertising, promotion, sale, distribution, import and export are subject to comprehensive regulation by the FDA and other regulatory agencies in the United States and by comparable authorities in other countries. Before we can commercialize any of our product candidates, we must obtain marketing approval. We have not received approval or clearance to market any of our

product candidates from regulatory authorities in any jurisdiction and it is possible that none of our product candidates or any product candidates we may seek to develop in the future will ever obtain regulatory approval or clearance. We have only limited experience in filing and supporting the applications necessary to gain regulatory approvals and may need to rely on third-party CROs and regulatory consultants to assist us in this process. Securing regulatory approval requires the submission of extensive preclinical and clinical data and supporting information to the various regulatory authorities for each therapeutic indication to establish the product candidate's safety and efficacy. Securing regulatory approval also requires the submission of information about the biologic manufacturing process to, and inspection of manufacturing facilities by, the relevant regulatory authority. Our product candidates may not be effective, may be only moderately effective or may prove to have undesirable or unintended side effects, toxicities or other characteristics that may preclude our obtaining marketing approval or prevent or limit commercial use. The FDA and other regulatory authorities have substantial discretion in the approval process, and determining when or whether regulatory approval will be obtained for a product candidate. Even if we believe the data collected from clinical trials are promising, such data may not be sufficient to support approval by the FDA or any other regulatory authority. In addition to the United States, we may seek regulatory approval to commercialize our product candidates in foreign countries. While the scope of regulatory approval is similar in many countries, to obtain separate regulatory approval in multiple countries will require us to comply with numerous and varying regulatory requirements of each such country or jurisdiction regarding safety and efficacy and governing, among other things, clinical trials and commercial sales, pricing and distribution, and we cannot predict success in any such jurisdictions.

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

- the FDA or comparable foreign regulatory authorities may disagree with the number, design or implementation of our clinical trials to support further development or approval;
- we may be unable to demonstrate to the satisfaction of the FDA or comparable foreign regulatory authorities that a product candidate is safe and effective for its proposed indication;
- the results of clinical trials may 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 foreign regulatory authorities;
- we may be unable to demonstrate that a product candidate's clinical and other benefits outweigh its safety risks;
- the FDA or comparable foreign regulatory authorities may disagree with our interpretation of data from preclinical studies or clinical trials;
- the FDA or comparable foreign regulatory authorities may disagree that we have provided sufficient safety data or adequately demonstrated clinical benefit in a patient population or subpopulation studied in the clinical trial;

- the data collected from clinical trials of our product candidates may not be sufficient to support the submission of a BLA or other submission or to obtain regulatory approval in the United States or elsewhere;

- the FDA or comparable foreign regulatory authority could require us to collect additional data or conduct additional clinical studies, for example, based on FDA feedback, we anticipate that to help inform the benefit-risk 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 effectiveness and safety of mavrilimumab beyond 26 weeks as well as evaluate its pharmacokinetic profile and the effectiveness of mavrilimumab at different doses;

- the FDA or comparable foreign regulatory authority could require us to conduct additional clinical studies to compare our product candidates to other therapies for the treatment of the same indication;

- the number of patients required for clinical trials of our product candidates may be larger than we anticipate, enrollment in these clinical trials may be slower than we anticipate, participants may drop out of these clinical trials at a higher rate than we anticipate or we may fail to recruit suitable patients to participate in a trial;

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

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

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

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

- the approval policies or regulations of the FDA or comparable foreign regulatory authorities may significantly change in a manner rendering our clinical data insufficient for approval.

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

If we experience delays in obtaining approval or if we fail to obtain approval of 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 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 licensed by the FDA. In addition, the approval of a biosimilar product may not be made effective by the FDA until 12 years from the date on which the reference product was first licensed. During this 12-year period of exclusivity, another company may still market a competing version of the reference product if the FDA approves a full BLA for the competing product containing the sponsor's own 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.

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

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

Even if we obtain marketing approval of our product candidates in a major pharmaceutical market such as the United States or the 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. Clinical trials conducted in one country may not be accepted by regulatory authorities in other countries, and regulatory approval in one country does not mean that regulatory approval will be obtained in any other country. Approval procedures vary among countries and can involve additional product testing and validation and additional administrative review periods. Seeking regulatory approvals in all markets may require additional preclinical studies or clinical trials, which would be costly and time consuming. Regulatory requirements can vary widely from country to country and could delay or prevent the introduction of our product candidates in those countries. Satisfying these and other regulatory requirements is costly, time consuming, uncertain and subject to unanticipated delays. In addition, our failure to obtain regulatory approval in any country may delay or have negative effects on the process for regulatory approval in other countries.

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

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

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

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

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

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

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

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 this designation, so 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 have obtained Fast Track Designation for one or more of our product candidates, we may not experience a faster development process, review or approval compared to non-expedited FDA review procedures. In addition, the FDA may withdraw Fast Track Designation if it believes that the designation is no longer supported. Fast Track Designation alone does not guarantee qualification for the FDA's priority review procedures.

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

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

Conducting clinical trials and preparing, and obtaining marketing approval for, a product candidate is a complicated process. Although members of our management team have participated in pivotal trials and obtained marketing approvals for product candidates in the past while employed at other companies, we as a company have not done so. As a result, such activities may require more time and cost more than we anticipate. Failure to successfully complete, or delays in, our global, pivotal Phase 3 clinical trial in rilonacept or any of our eventual other pivotal trials or related regulatory submissions would prevent us from, or delay us in, obtaining regulatory approval for, or clearance of, our product candidates. In addition, it is possible that the FDA or other government agencies may refuse to accept for substantive review any regulatory submissions that we submit for our product candidates or may conclude after review of our applications that they are insufficient to obtain marketing approval or clearance of our product candidates. If the FDA or other government agencies do not accept our applications or issue marketing authorizations for our product candidates, they may require that we conduct additional clinical, preclinical or manufacturing validation trials and submit that data before they will reconsider our applications. Depending on the extent of these or any other required trials, approval or receipt of any marketing authorization may be delayed by several years or may require us to expend more resources than we have available. It is also possible that additional trials, if performed and completed, may not be considered sufficient by the FDA or other government agencies to approve or grant marketing authorizations. Any delay in obtaining, or an inability to obtain, marketing approvals would prevent us from commercializing our product candidates, generating revenues and achieving and sustaining profitability. If any of these outcomes occur, we may be forced to modify or cease our development efforts for our product candidates, which could significantly harm our business.

Changes in funding for the FDA and other government agencies could hinder their ability to hire and retain key leadership and other personnel, or otherwise prevent new products and services from being developed or commercialized in a timely manner, which could negatively impact our business.

The ability of the FDA or other government agencies to review and approve new products can be affected by a variety of factors, including government budget and funding levels, ability to hire and retain key personnel and accept the payment of user fees, and statutory, regulatory, and policy changes. Average review times at the agency have fluctuated in recent years as a result. In addition, government funding of other government agencies that fund research and development activities is subject to the political process, which is inherently fluid and unpredictable.

Disruptions at the FDA and other government agencies may also slow the time necessary for new drugs to be reviewed 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. If a prolonged government shutdown occurs, it could significantly impact the ability of the FDA 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 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 manufacturing facility to produce drug substance to support certain preclinical and early clinical development for our product candidates, we rely, and expect to continue to rely, on third parties for the manufacture of our product candidates for the majority of our development efforts, as well as for the potential commercial manufacture of our product candidates, if approved. We rely on these third parties to develop the processes necessary to produce our product candidates at sufficient quality and quantity to support our development and commercialization efforts. Our reliance increases the risk that we will have insufficient quantities of our product candidates or that our product candidates are not produced at an acceptable cost or quality, which could delay, prevent or impair our development or commercialization efforts.

For example, we have a contract with Regeneron to produce rilonacept on an exclusive basis for a period of time. Although Regeneron has produced rilonacept for commercial use for over ten years, the FDA or another applicable foreign regulatory agency might reevaluate rilonacept's current manufacturing process or route of administration in connection with evaluating whether to approve rilonacept for a new indication, such as recurrent pericarditis. We also have CMOs manufacture KPL-716 drug substance and drug product and entered into an agreement with a CMO to produce mavrilimumab beyond our current inventory. While we have transferred the technology to manufacture mavrilimumab to the CMO, the CMO may be required to adopt different manufacturing protocols or processes. The CMO will need to produce mavrilimumab using different media and feed compared to the processes that were used by MedImmune to develop our existing inventory. We cannot provide any assurance that the technology transfer was successful, or that the process development or the CMO will be successful in producing mavrilimumab in sufficient quantities or of acceptable quality, if at all. In addition, we contract with CMOs in connection with certain production and testing of our preclinical product candidates. While we have built a manufacturing facility to support certain preclinical and early clinical development for our product candidates, we and our CMOs may not be able to produce sufficient quantities of these product candidates or produce them at an acceptable quality, which could delay, prevent or impair our development or commercialization efforts and increase costs.

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. 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 others, 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 a comparable foreign regulatory authority does not approve these facilities for the manufacture of our product candidates or if it withdraws any such approval in the future, we may need to find alternative manufacturing facilities, which would significantly impact our ability to develop, obtain regulatory approval for or market our product candidates, if approved. Further, our failure, or the failure of our third-party manufacturers, to comply with applicable regulations could result in sanctions being imposed on us, including clinical holds, fines, injunctions, civil penalties, delays, suspension or withdrawal of approvals, license revocation, seizures or recalls of product candidates, if approved, operating restrictions and criminal prosecutions, any of which could significantly and adversely affect our business and supplies of our product candidates.

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

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

Our product candidates may compete with other product candidates and approved products for access to manufacturing facilities. There are a limited number of manufacturers that operate under cGMP regulations and that might be capable of manufacturing for us.

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

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

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

We built a manufacturing facility to support the early development of our product candidates, and we may be unsuccessful in manufacturing product candidates in a timely, economic or compliant manner, which could delay or prevent the commencement of our planned preclinical and early clinical studies for these product candidates.

We built a manufacturing facility to support preclinical and early clinical studies for our product candidates. We may not be able to continually manufacture our product candidates economically or in compliance with cGMPs and other regulatory requirements, or at all, and we may not be able to build or procure additional capacity in the required timeframe to meet our estimated timelines to commence our studies. As a result, our business, prospects, operating results, and financial condition could be materially harmed.

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

Our research and development activities and our third-party manufacturers' and suppliers' activities involve the controlled storage, use and disposal of hazardous materials owned by us, including the components of our product candidates and other hazardous compounds. We and our manufacturers and suppliers are subject to laws and regulations

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

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

The manufacture of our product candidates is highly regulated, complex and difficult, requiring a multi-step and controlled process, and even minor problems or deviations could result in the product candidates being out-of-spec, failed batches or other failures, such as defective products or manufacturing failures. We have limited experience overseeing the manufacturing processes of mavrilimumab, KPL-716, and our preclinical product candidates 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, including natural disasters, accidents, labor disputes, acts of terrorism or war. The occurrence of any such event could adversely affect our ability to satisfy the required supply for any of our product candidates 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 could result in a product shortage of clinical or commercial requirements, withdrawal of our product candidates or any approved products, shipment delays, lot failures or recalls. We may also have to write off inventory and incur other charges and expenses for products that fail to meet specifications, undertake costly remediation efforts or seek more costly manufacturing

alternatives. Such manufacturing issues could increase our cost of goods, cause us to lose potential revenue, reduce our potential profitability or damage our reputation.

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

The drug substance and drug product used in rilonacept, mavrilimumab, KPL-716 and our preclinical candidates are supplied to us from single-source suppliers. For example, Regeneron has a contractual right to be our sole source manufacturer of the product unless they have a persistent failure to satisfy our supply needs. Our ability to successfully develop our product candidates, and to ultimately supply our commercial products in quantities sufficient to meet the market demand, depends in part on our ability to obtain the 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 KPL-716 in the quantities that we believe would be required to meet anticipated market demand, our third-party manufacturers may need to increase manufacturing capacity and, in some cases, alternative sources of commercial supply may need to be secured, which could involve significant challenges and may require additional regulatory approvals. In addition, the development of commercial-scale manufacturing capabilities may require us and our third-party manufacturers to invest substantial additional funds and hire and retain the technical personnel who have the necessary manufacturing experience. Neither we nor our third-party manufacturers may successfully complete any required increase to existing manufacturing capacity in a timely manner, or at all.

Moreover, if there is a disruption to one or more of our third-party manufacturers' or suppliers' relevant operations the supply of the related product candidate will be delayed until such manufacturer or supplier restores the affected facilities or we or they procure alternative manufacturing facilities or sources of supply. Our ability to progress our preclinical and clinical programs could be materially and adversely impacted if any of the third-party suppliers upon which we rely 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, may not be accomplished quickly and can take several years, if at all. Furthermore, despite our efforts, we may be unable to procure a replacement supplier or do so on commercially reasonable terms, which could have a material adverse impact upon our business. If we are able to find a replacement supplier, such replacement supplier would need to be qualified and may require additional regulatory approval, which could result in further delay. While we seek to maintain adequate inventory of the 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 at acceptable prices in a timely manner could impede, delay, limit or prevent our development efforts, which could harm our business, results of operations, financial condition and prospects.

Certain of the materials required in the manufacture and the formulation of our product candidates are derived from biological sources. Such materials are difficult to procure and may be subject to contamination or recall. Access to and supply of sufficient quantities of raw materials which meet the technical specifications for the production process is

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

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

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

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

Although we have and intend to continue to design the clinical trials for our product candidates, CROs will conduct and oversee all of the clinical trials together with the various clinical trial sites that we engage to conduct the studies. As a result, many important aspects of our development programs, including their conduct and timing, will be outside of our direct control. Our reliance on third parties to conduct future clinical trials will also result in less direct control over the management of data developed through clinical trials than would be the case if we were relying entirely

upon our own staff. Communicating with outside parties can also be challenging, potentially leading to mistakes as well as difficulties in coordinating activities. Outside parties may:

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

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

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

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

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

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

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

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

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

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

While we are not aware of any therapies currently approved or actively continuing clinical trials in recurrent pericarditis, there is one product that modulates the signaling of IL-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 is also another therapy which modulates IL-1α in clinical development for diseases other than recurrent pericarditis from XBiotech Inc. We expect mavrilimumab, if approved, to experience competitive pressure from tocilizumab (ACTEMRA), marked 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 perusing GCA in clinical trials that could decide in the future to engage in development of therapies for GCA, including GlaxoSmithKline plc, Izana Bioscience and Humanigen, Inc. Multiple therapies are in development for prurigo nodularis and any that receive FDA approval for this indication will be likely competitors to KPL-716. These products include nemolizumab, serlopitant and nalbuphine ER. 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. We may seek to acquire businesses or undertake business combinations, collaborations, or other strategic transactions but we may not realize the intended benefits of such transactions.

We have acquired or in-licensed our existing product candidates, and 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 transactions 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. We cannot be certain that we will be successful in such efforts 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;
- 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 or strategic transactions, or our growth strategy or strategic acquisitions may not deliver the anticipated results.

The consummation or performance of any acquisition, business combination, collaboration or other strategic transaction we may undertake in furtherance of our growth 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.

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

We may seek collaboration or licensing arrangements for the development, or potentially for the commercialization, of certain 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 arrangements. In addition, we may seek to jointly develop or commercialize one or more of our product candidates. We will face, to the extent that we decide to enter into such arrangements, significant competition in seeking appropriate collaborators. Moreover, these arrangements are complex and time consuming to negotiate, document, implement and maintain. We may not be successful in our efforts to establish and implement collaborations or other alternative arrangements should we so chose to enter into such arrangements. The terms of any collaborations or other arrangements that we may establish may not be favorable to us. In addition, our right to grant a sublicense of intellectual property licensed to us under certain of our current agreements requires the consent of the applicable licensor.

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

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

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

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

We have a limited operating history and are highly dependent on the research and development, clinical, commercial and business development expertise of members of our executive and senior management teams, as well as the other principal members of our management, scientific and clinical teams. Although we have entered into employment agreements with our executive officers and 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. 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 and expand our company, and we may encounter difficulties in managing this development and expansion, which could disrupt our operations.

We expect to continue to increase our number of employees and the scope of our operations. To manage our anticipated development and expansion, we must continue to implement and improve our managerial, operational and financial systems, expand our facilities and continue to recruit and train additional qualified personnel. Also, our management may need to divert a disproportionate amount of its attention away from its day-to-day activities and devote a substantial amount of time to managing these development activities. Due to our limited resources, certain employees may need to perform activities that are beyond their regular scope of work, and we may not be able to effectively manage the expansion of our operations or recruit and train additional qualified personnel. This may result in weaknesses in our infrastructure, give rise to operational mistakes, loss of business opportunities, loss of employees and reduced productivity among remaining employees. The physical expansion of our operations may lead to significant costs and may divert financial resources from other projects, such as the development of our product candidates. If our management is unable to effectively manage our expected development and expansion, our expenses may increase more than expected, our ability to generate revenue could be reduced and we may not be able to implement our business strategy. Our future financial performance and our ability to commercialize our product candidates, if approved, and to compete effectively will depend, in part, on our ability to effectively manage the future development and expansion of our company.

Risks Related to Intellectual Property

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

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

We acquire, in-license and file patent applications directed to our product candidates in an effort to establish intellectual property positions directed to their compositions of matter as well as uses of these product candidates in the treatment of diseases. Our intellectual property includes patents and patent applications that we own as well as patents and patent applications that we in-license. For example, we have a field-specific exclusive license under a license agreement with Regeneron to patent applications and patents relating to rilonacept, an exclusive license under a license agreement with MedImmune, or the MedImmune Agreement, to patent applications and patents relating to mavrilimumab, and an exclusive license under our license agreement with Novo Nordisk to patent applications, patents relating to KPL-045 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, KPL-716 or our other product candidates. In addition, the laws of foreign countries may not protect our rights to the same extent as the laws of the United States. Furthermore, patents have a limited lifespan. In the United States, the natural expiration of a patent is generally 20 years after it is filed. Various extensions and adjustments may be available; however, the life of a patent, and the protection it affords, is limited. The actual protection afforded by a patent varies on a product-by-product basis, from country to country and depends upon many factors, including the type of patent, the scope of its coverage, the availability of regulatory-related extensions, the availability of legal remedies in a particular country and the validity and enforceability of the patent.

Patents may be eligible for limited patent term extension in the United States under the Drug Price Competition and Patent Term Restoration Act of 1984, referred to as the Hatch-Waxman Act. Similar patent extensions exist in the 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 expires in 2019 in the United States, not including patent term adjustment, and in 2023 in Europe, not including any patent term extensions, and the patents covering mavrilimumab as a composition of matter have a term that expires in 2027 in the United States, not including any patent term adjustments or extensions, and in 2027 in Europe, not including any patent term extensions. As a result, our owned and in-licensed patent portfolio may not provide us with adequate and continuing patent protection sufficient to exclude others from commercializing products similar or identical to our product candidates. In such cases, we expect to rely on regulatory exclusivity for our product candidates, such as orphan drug exclusivity, which generally grants seven years of marketing exclusivity under the Federal Food, Drug, and Cosmetic Act, and up to ten years of marketing exclusivity in Europe. While, we may pursue orphan drug designation for our product candidates in the United States, we may not be successful in obtaining such designation or we may not be able to maintain the benefits of the designation. Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug

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

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

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

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

also become involved in similar opposition proceedings in the European Patent Office or similar offices in other jurisdictions regarding our intellectual property rights with respect to our products and technology.

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

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

In addition, we may in the future be subject to claims by our 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, KPL-716, KPL-404 and KPL-045. In September 2017, we entered into a license agreement

with Regeneron to obtain an exclusive license under certain intellectual property rights controlled by Regeneron to develop and commercialize rilonacept. In December 2017, we entered into a license agreement with MedImmune to obtain exclusive worldwide rights to research, develop, manufacture, market and sell mavrilimumab and any other products covered by the licensed patent rights. In September 2016, pursuant to an asset purchase agreement with Biogen, or the Biogen Agreement, we acquired all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716, including patents and other intellectual property rights, clinical data, know-how and inventory. In connection with our acquisition of Primatope in March 2019, we acquired a license with Beth Israel Deaconess Medical Center for certain patent applications and patents related to KPL-404. In August 2017, we licensed KPL-045 from Novo Nordisk. 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 interleukin-1 receptor, and CAPS. Regeneron may also develop rilonacept in fields to which we have licensed the rights, but we retain the commercial benefit related to that development upon approval of rilonacept in any field that we have licensed. We and Regeneron communicate with each other concerning our related development activities, and we have approval rights over Regeneron's development in the fields that we have licensed, including pericarditis. Outside of the United States and Japan, Regeneron has granted a third-party licensee the right to develop and commercialize rilonacept in CAPS and certain periodic fever syndromes. 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 OSM receptor (other than for the development and commercialization of products that are the subject of the Biogen Agreement). This exclusivity obligation runs from the earlier of the eighth anniversary of the agreement or the first commercial sale of a product that is the subject of the agreement.

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

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

Given the vast number of patents in our field of technology, we cannot be certain that we do not infringe existing patents or that we will not infringe patents that may be granted in the future. Many companies have filed, and continue to file, patent applications related to immunomodulation. Some of these patent applications have already been allowed or issued, and others may issue in the future. For example, we are aware of third-party patents that contain claims potentially relevant to certain therapeutic uses of mavrilimumab and KPL-716. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to mavrilimumab and KPL-716 would be found to infringe any valid claim of these patents. While we may decide to initiate proceedings to challenge the validity of these or other patents in the future, we may be unsuccessful, and courts or patent offices in the United States and abroad could uphold the validity of any such patent. If we were to challenge the validity of any issued 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 foreign patent lawsuit, alleging our infringement of a competitor's patents, we could be prevented from marketing our products in one or more foreign countries. Any of these outcomes would have a materially adverse effect on our business.

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

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

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

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

The USPTO and various foreign governmental patent agencies require compliance with a number of procedural, documentary, fee payment and other similar provisions during the patent application process. In addition, periodic maintenance fees on issued patents often must be paid to the USPTO and foreign patent agencies over the lifetime of the patent. While an unintentional lapse can in many cases be cured by payment of a late fee or by other means in accordance with the applicable rules, there are situations in which noncompliance can result in abandonment or lapse of the patent or patent application, resulting in partial or complete loss of patent rights in the relevant jurisdiction. Non-compliance events that could result in abandonment or lapse of a patent or patent application include, but are not limited to, failure to respond to official actions within prescribed time limits, non-payment of fees and failure to properly

legalize and submit formal documents. If we or our licensors fail to maintain the patents and patent applications covering our products or technologies, we may not be able to stop a competitor from marketing products that are the same as or similar to our product candidates, which would have a material adverse effect on our business. In addition, if we fail to apply for applicable patent term extensions or adjustments, we will have a more limited time during which we can enforce our granted patents. In addition, if we are responsible for patent prosecution and maintenance of patent rights in-licensed to us, any of the foregoing could expose us to liability to the applicable patent owner.

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

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

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

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

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

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

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

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

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

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

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

information to compete with us. We may in the future rely on trade secret protection, which would be subject to the risks identified above with respect to confidential information.

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

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

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

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

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

Risks Related to Commercialization

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

The precise incidence and prevalence for all the conditions we aim to address with our programs are unknown. Our projections of both the number of people who have these diseases, as well as the subset of people with these diseases who have the potential to benefit from treatment with our product candidates, are based on our extrapolation

from available population data and estimates. These estimates have been derived from a variety of sources, including the scientific literature, surveys of clinics, patient foundations, pharmacy claims analyses, or market research, and may prove to be incorrect. Further, new trials and therapeutic options may change the estimated incidence or prevalence of these diseases. The number of patients may turn out to be lower than expected.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

While in some markets, there is a single payor, in other markets there are multiple payors that can have different ways of assessing prescription drugs. To commercialize our product candidates successfully, we will be required to have sufficient expertise and resources to execute on the respective product candidate's coverage and reimbursement strategy, which we cannot be certain we will be able to do.

In the interest of cost-containment, government authorities, private health insurers and other third-party payors have attempted to control costs by delaying the time to reimbursement, and by limiting the breadth of coverage and the amount of reimbursement for particular products, both 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 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, thereby impacting he progression to a price negotiation. The potential of third-party payors to introduce more rigorous value/benefit assessment processes and criteria could have a negative impact on our ability to commercialize our product candidates successfully.

Third-party payors are also introducing more challenging price negotiation tactics, including in re-visiting established coverage and reimbursement in cases when new competitors, including brands, generics and biosimilars enter the market. It is possible that a third-party payor may consider our product candidates as substitutable and only

offer to cover the cost of the alternative product. Even if we show improved efficacy or improved convenience of administration with our product candidates, pricing of competitive products may limit the amount we will be able to charge for our product candidates. Third-party payors may deny or revoke the reimbursement status of a given product or establish prices for new or existing marketed products at levels that are too low to enable us to realize an appropriate return on our investment in our product candidates. In some cases, when new competitor generic and biosimilar products enter the market, there are mandatory price reductions for the innovator compound, in other cases, payors employ "therapeutic category" price referencing and seek to lower the reimbursement levels for all treatment in the respective therapeutic category. In other cases, new competitor brand drugs can trigger therapeutic category reviews in the interest of modifying coverage and or reimbursement levels. The potential of third-party payors to introduce more challenging price negotiation tactics could have a negative impact on our ability to commercialize our product candidates successfully.

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

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. This is a tactic that is in a state of constant potential change as countries seek to establish "reference baskets" of countries that will enable lower pricing. International reference pricing adds to the regional impact of price cuts in individual countries and expansion of international reference pricing, including into the United States, presents a material risk to our ability to achieve satisfactory coverage and reimbursement.

Drug importation and cross-border trade, both sanctioned and unsanctioned, occurs when a pharmaceutical product from a lower cost market 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, increasingly high barriers are being erected to the entry of new products.

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

Our future profitability may depend, in part, on our ability to commercialize our product candidates in foreign markets for which we may rely on collaborations with third parties. For example, we are enrolling a global, pivotal Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis. Although we do not have immediate plans to pursue the commercialization of rilonacept for recurrent pericarditis outside of the United States, we are evaluating the opportunities for the development and commercialization of our product candidates in certain foreign markets. We are not permitted to market or promote any of our product candidates before we receive regulatory approval from the applicable regulatory authority in that foreign market, and we may never receive such regulatory approval for any of our product candidates. To obtain separate regulatory approval in many other countries we must comply with numerous and varying regulatory requirements of such countries regarding safety and efficacy and governing, among other things, clinical trials and commercial sales, pricing and distribution of our product candidates, and we cannot predict success in these jurisdictions. If we obtain approval of our product candidates and ultimately commercialize our product candidates in foreign markets, we would be subject to additional risks and uncertainties, including:

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

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

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

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

If the FDA or a comparable foreign regulatory authority approves any of our product candidates, it 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, including both federal and state requirements in the United States and requirements of comparable foreign regulatory authorities.

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

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

We will have to comply with requirements concerning advertising and promotion for our products. Promotional communications with respect to prescription drugs and biologics are subject to a variety of legal and regulatory restrictions and must be consistent with the information in the product's approved label. As such, we may not promote our products for indications or uses for which they do not have approval. Even our current clinical and medical affairs activities are subject to certain ongoing regulatory requirements concerning appropriate exchange of medical and scientific information.

The holder of an approved BLA or MAA must submit new or supplemental applications and obtain approval for certain changes to the approved product, product labeling or manufacturing process. We could also be asked to conduct post-marketing clinical trials to verify the safety and efficacy of our products in general or in specific patient subsets. If original marketing approval were obtained via the accelerated approval pathway, we could be required to conduct a successful post-marketing clinical trial to confirm clinical benefit for our products. An unsuccessful post-marketing trial or failure to complete such a trial could result in the withdrawal of marketing approval. The FDA also may place other conditions on approvals including the requirement for a REMS, to assure the safe use of the product. If the FDA concludes a REMS is needed, the sponsor of the BLA must submit a proposed REMS before it can obtain approval. A REMS could include medication guides, physician communication plans or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools.

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

- issue warning letters;
- impose civil or criminal penalties;

- suspend or withdraw regulatory approval;
- suspend any of our ongoing clinical trials;
- refuse to approve pending applications or supplements to approved applications submitted by us;
- impose restrictions on our operations, including closing our CMOs' facilities; or
- seize or detain products, or require a product recall.

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

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

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

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

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

regulations that may constrain the business or financial arrangements and relationships through which we market, sell and distribute our product candidates for which we obtain marketing approval.

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

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

- the U.S. federal False Claims Act and civil monetary penalties laws, which, among other things, impose criminal and civil penalties, including through civil whistleblower or *qui tam* actions, against individuals or entities for knowingly presenting, or causing to be presented, to the federal government, claims for payment that are false or fraudulent or making a false statement to avoid, decrease or conceal an obligation to pay money to the federal government. In addition, the government may assert that a claim including items and services resulting from a violation of the federal Anti-Kickback Statute constitutes a false 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 and teaching hospitals (and additional categories of healthcare practitioners beginning with reports due on or after January 1, 2022) and 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. 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

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. The current Presidential Administration and U.S. Congress have attempted and will likely continue to seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Most recently, the Tax Cuts and Jobs Act was enacted, which, among other things, removes penalties for not complying with the Affordable Care Act's individual mandate to carry health insurance. A recent federal district court ruling struck down the Affordable Care Act in its entirety. This decision means numerous reforms enacted as part of the Affordable Care Act, but not specifically related to health insurance, such as the BPCIA, are invalid as well. While the presidential administration and The Centers for Medicare & Medicaid Services, or CMS, have both stated that the ruling will have no immediate effect, it is unclear how this decision, subsequent appeals, if any, and other efforts to repeal and replace the Affordable Care Act will impact the Affordable Care Act and our business.

In addition, other legislative changes have been proposed and adopted in the United States since the Affordable Care Act was enacted. The Budget Control Act of 2011, among other things, led to aggregate reductions of Medicare payments to providers of 2% per fiscal year. These reductions went into effect on April 1, 2013 and, due to subsequent legislative amendments to the statute, will remain in effect through 2027 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, and in October 2018, CMS proposed a new rule that would require direct-to-consumer television advertisements of prescription drugs and biological products, for which payment is available through or under Medicare or Medicaid, to include in the advertisement the wholesale acquisition cost, or list price, of that drug or biological product. Moreover, payment methodologies may be subject to changes in healthcare legislation and regulatory initiatives. We expect that additional U.S. federal healthcare reform measures will be adopted in the future, any of which could limit the amounts that the U.S. federal government will pay for healthcare products and services, which could result in reduced demand for our product candidates or additional pricing pressures.

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

In the EU, similar political, economic and regulatory developments may affect our ability to profitably commercialize our product candidates, if approved. In addition to continuing pressure on prices and cost containment measures, legislative developments at the EU or member state level may result in significant additional requirements or obstacles that may increase our operating costs. The delivery of healthcare in the EU, including the establishment and operation of health services and the pricing and reimbursement of medicines, is almost exclusively a matter for national, rather than EU, law and policy. National governments and health service providers have different priorities and approaches to the delivery of health care and the pricing and reimbursement of products in that context. In general, however, the healthcare budgetary constraints in most EU member states have resulted in restrictions on the pricing and reimbursement of medicines by relevant health service providers. Coupled with ever-increasing EU and national regulatory burdens on those wishing to develop and market products, this could prevent or delay marketing approval of our product candidates, restrict or regulate post-approval activities and affect our ability to commercialize our product candidates, if approved.

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

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

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

Our results of operations could be adversely affected by general conditions in the global economy and in the global financial markets. For example, the global financial crisis caused extreme volatility and disruptions in the capital

and credit markets. A severe or prolonged economic downturn, such as the global financial crisis, could result in a variety of risks to our business, including weakened demand for our product candidates and our ability to raise additional capital when needed on acceptable terms, if at all. A weak or declining economy could also strain our suppliers, possibly resulting in supply disruption. 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 foreign operations;
- complexities associated with managing multiple payor reimbursement regimes, government payors or patient self-pay systems;
- limits in our ability to penetrate international markets;
- financial risks, such as longer payment cycles, difficulty collecting accounts receivable, the impact of local and regional financial crises on demand and payment for our products and exposure to foreign currency exchange rate fluctuations;
- natural disasters, political and economic instability, including wars, terrorism, political unrest, outbreak of disease and boycotts;
- curtailment of trade, and other business restrictions;
- certain expenses including, among others, expenses for travel, translation and insurance; and
- regulatory and compliance risks that relate to maintaining accurate information and control over sales and activities that may fall within the purview of the U.S. Foreign Corrupt Practices Act, its books and records provisions or its anti-bribery provisions.

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

Our internal 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 product candidates' development programs or loss of other assets, including funds.

Despite the implementation of security measures, our internal technology systems and those of our third-party CMOs, CROs and other contractors, consultants and service providers are vulnerable to damage from viruses, unauthorized access and attacks, theft, natural disasters, terrorism, war and telecommunication and electrical failures. If such an event were to occur and cause interruptions in our business and operations or those of our third-party CMOs, CROs and other contractors, consultants and service providers, it could result in a material disruption of our or such third-party's business or operations and our product candidates' development programs or loss of other assets, including

funds. For example, the loss of clinical trial data for our product candidates could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any disruption or security breach results in a loss of or damage to our data or applications or other data or applications relating to our technology or product candidates, or inappropriate disclosure or theft of confidential or proprietary information, we could incur liabilities and the further development of our product candidates could be delayed.

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

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

Our clinical trial programs outside the United States may implicate international data protection laws, including the EU Data Protection Directive and legislation of the EU member states implementing it. Our activities outside the United States impose additional compliance requirements and generate additional risks of enforcement for noncompliance. Failure by our CROs and other third-party contractors to comply with the strict rules on the transfer of personal data outside of the EU into the United States may result in the imposition of criminal and administrative sanctions on such collaborators, which could adversely affect our business.

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

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

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

In May 2018, a new privacy framework, the General Data Protection Regulation, or the GDPR, took effect in the EU and became binding across all EU member states. The GDPR is in the process of taking effect in the EEA. The GDPR imposes several stringent requirements for controllers and processors of personal data, particularly with respect to clinical trials. The GDPR provides that EU member states may make their own further laws and regulations limiting the processing of genetic, biometric or health data, which could limit our ability to use and share personal data or could cause our costs to increase and harm our business and financial condition. In addition, the GDPR increases the scrutiny

that clinical trial sites located in the EEA should apply to transfers of personal data from such sites to countries that are considered to lack an adequate level of data protection, such as the United States. There are currently a number of legal challenges to the validity of EU mechanisms for adequate data transfers (such as the commonly-used EU-Commission-approved model clauses) or review of these mechanisms (such as the U.S. Privacy Shield), and our business could be impacted by changes in law as a result of a future review of these transfer mechanisms by EU regulators under the GDPR, as well as current challenges to these mechanisms in the EU courts. The GDPR imposes substantial fines for breaches of data protection requirements, which can be up to four percent of global revenue for the preceding financial year or €20 million, whichever is greater, and it also confers a private right of action on data subjects for breaches of data protection requirements. Compliance with EU data protection law is a rigorous and time-intensive process that may increase our cost of doing business, and despite those efforts, there is a risk that we may be subject to fines and penalties, litigation and reputational harm in connection with our European activities.

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. 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.

We have adopted a code of conduct applicable to all of our employees, but it is not always possible to identify and deter misconduct by employees and other third parties. 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 managed by Baker Brothers, and Class B1 common shares, all of which shares are held by entities managed by Baker Brothers, means that such persons are, and such entities may in the future be, able to influence or control certain matters submitted to our shareholders for approval; and such concentration and conversion rights and resulting concentration of control may have an adverse effect on the price of our Class A common shares and may result in our Class A common shares being undervalued.

Each Class A common share is entitled to one vote per Class A common share and each Class B common share is entitled to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares have no voting rights. As a result, all matters submitted to our shareholders are decided by the vote of holders of our Class A common shares and Class B common shares. As a result of the multi-class voting structure of our common shares, the holders of our Class B common shares, which consist primarily of our executive officers and certain other members of our senior management, collectively control over a majority of the combined voting power of our common shares and therefore are able to control the outcome of certain matters submitted to our shareholders for approval. As of June 30, 2019, the holders of Class A common shares accounted for approximately 29% of our aggregate voting power and the holders of Class B common shares accounted for approximately 71% 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 66% of our aggregate voting power as of June 30, 2019 and have the ability to control the outcome of certain matters submitted to our shareholders for approval.

However, this percentage may change depending on any conversion of our Class B common shares, Class A1 common shares or Class B1 common shares. Each holder of our Class B common shares has the ability to convert any portion of its Class B common shares into Class B1 common shares or Class A common shares at any time with advance notice to us. Each holder of our Class B1 common shares has the ability to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time with advance notice to us, and each holder of our Class A1 common shares has the ability to convert any portion of its Class A1 common shares into Class A common shares at any time with advance notice to us. Our Class A1 common shares and Class B1 common shares cannot be converted if, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of our issued and outstanding Class A common shares unless such holder provides us with 61-days' prior notice that it intends to increase, decrease or waive such threshold upon conversion. As of June 30, 2019, entities managed by Baker Brothers 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 74% of our aggregate voting power.

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

This concentrated control limits certain shareholders' ability to influence corporate matters and may have an adverse effect on the price of our Class A common shares, including our Class A common shares being undervalued. Holders of our Class B common shares collectively control our management and affairs and are able to influence or control the outcome of certain matters submitted to our shareholders for approval, including the election of directors.

Due to the conversion rights of the holders of our Class A1 and B1 common shares, entities affiliated with the Baker Brothers could significantly increase their voting control of us, which could result in their ability to significantly influence or control matters submitted to our shareholders for approval. These holders may have interests, with respect to their investment, that are different from our other shareholders. In addition, this concentration of control might adversely affect certain corporate actions that our other shareholders may view as beneficial, for example, by:

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

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

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

- the results of clinical trials for our product candidates;
- delays in the commencement, enrollment and the ultimate completion of clinical trials;
- the results and potential impact of competitive products or technologies;
- our ability to manufacture and successfully produce our product candidates;
- actual or anticipated changes in estimates as to financial results, development timelines or recommendations by securities analysts;
- the level of expenses related to any of our product candidates or clinical development programs;
- variations in our financial results or those of companies that are perceived to be similar to us;
- financing or other corporate transactions, or our inability to obtain additional funding;
- failure to meet or exceed the expectations of the investment community;
- regulatory or legal developments in the United States and other countries;
- the recruitment or departure of key personnel;
- developments or disputes concerning patent applications, issued patents or other proprietary rights;
- the results of our efforts to discover, develop, acquire or in-license additional product candidates;
- changes in the structure of healthcare payment systems;
- market conditions in the pharmaceutical and biotechnology sectors;

- general economic, industry and market conditions;
- changes in voting control of our executive officers and certain other members of our senior management or affiliates who hold our shares; and
- the other factors described in this "Risk Factors" section.

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

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

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

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

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

As of June 30, 2019, upon such transfers or conversions up to approximately 35.7 million of additional Class A common shares would be issuable and eligible for resale in the public market to the extent permitted by the provisions of Rule 144 promulgated under the Securities Act of 1933, as amended, or the Securities Act and such rule, Rule 144. In addition, as of June 30, 2019, there were approximately 7.4 million Class A common shares subject to outstanding options under our equity incentive plans that may become eligible for sale in the public market to the extent permitted by the provisions of applicable vesting schedules and Rule 144 and Rule 701 under the Securities Act.

Over a majority of our common shares are held by our executive officers and other members of our senior management together with funds affiliated with certain of our directors. As of June 30, 2019, on an as-converted basis, these shareholders collectively held approximately 33.8 million of our Class A common shares. If any of these shareholders sell, convert or transfer, or indicate an intention to sell, convert or transfer, a substantial amount of their common shares (after certain restrictions on conversion or resale lapse), the market price of our Class A common shares could decline.

Further, as of June 30, 2019, (i) holders of approximately 35.0 million Class A common shares, including Class A common shares issuable upon the conversion of our Class B, Class A1 and Class B1 common shares and upon the exercise of certain rights to acquire Class A common shares, or collectively, registerable securities, are entitled to certain rights with respect to the registration of these shares under the Securities Act pursuant to our amended and restated investor rights agreement, or the investors rights agreement and (ii) we had registered 28,882,977 Class A common shares (inclusive of 3,000,000 Class A common shares acquired by certain of these holders in our IPO) on an as converted basis from the registrable securities held by certain of these holders pursuant to the investors rights agreement, which are freely tradable without restriction under the Securities Act, to the extent permitted by Rule 144 under the Securities Act.

If any of these Class A common shares are sold, or if it is perceived that they will be sold, in the public market, the market price of our Class A common shares could decline.

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

We will need additional capital in the future to continue our planned operations. To the extent we raise additional capital by issuing additional Class A common shares, Class B common shares, Class A1 common shares, Class B1 common shares or other equity securities, our shareholders may experience substantial dilution. We may sell common shares, convertible securities or other equity securities in one or more transactions at prices and in a manner we determine from time to time under our shelf registration statement or otherwise. If we sell common shares, convertible securities or other equity securities in more than one transaction, investors may be materially diluted by subsequent sales. These sales may also result in material dilution to our existing shareholders, and new investors could gain rights superior to our existing shareholders.

In addition, the consummation or performance of any acquisition, business combination, collaboration or other strategic transaction we may undertake in furtherance of our growth strategy may cause dilution to our existing shareholders if we issue equity securities for all or a portion of the consideration. For example, we acquired the issued and outstanding equity securities of Primatope in exchange for upfront consideration of $10.0 million paid at closing in March 2019 as well as milestone payments of $5.0 million, which had been achieved and paid as of the closing date, and $3.0 million which was paid in June 2019, all of which paid in a combination of cash and our Class A common shares (inclusive of escrow and holdback amounts) in accordance with the terms and conditions of our stock purchase option agreement with Primatope.

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

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

- the last day of the fiscal year in which we have total annual gross revenues of $1.07 billion or more;
- the last day of the fiscal year following the fifth anniversary of the date of the completion of our IPO;
- the date on which we have issued more than $1 billion in nonconvertible debt during the previous three years; or

- the date on which we are deemed to be a large accelerated filer under the rules of the SEC, which means the market value of our voting and non-voting common shares that is held by non-affiliates exceeds $700 million as of the prior June 30th.

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

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

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

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

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

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

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

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

As a public company, we incur significant legal, accounting and other expenses. In addition, the Sarbanes-Oxley Act of 2002 and rules subsequently implemented by the SEC and The Nasdaq Global Select Market, or Nasdaq, where our Class A common shares are listed, impose various requirements on public companies, including establishment and maintenance of effective disclosure and financial controls and corporate governance practices. Our management and other personnel devote a substantial amount of time to these compliance initiatives. Moreover, these rules and regulations increase our legal and financial compliance costs and make some activities more time-consuming and costly, and will increase after we are no longer an emerging growth company and a smaller reporting company. For example, these rules and regulations may make it more difficult and more expensive for us to obtain director and officer liability insurance.

Pursuant to Section 404, we are required to furnish a report by our management on our internal control over financial reporting, including an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. However, while we remain an emerging growth company, we are not required to include an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. To maintain compliance with Section 404 within the prescribed period, we engage in a process to document and evaluate our internal control over financial reporting, which is both costly and challenging. In this regard, we will need to continue to dedicate internal resources, engage outside consultants and refine and revise a detailed work plan to assess and document the adequacy of internal control over financial reporting, continue steps to improve control processes as appropriate, validate through testing that controls are functioning as documented and implement a continuous reporting and improvement process for internal control over financial reporting. Despite our efforts, there is a risk that neither we nor our independent registered public accounting firm will be able to conclude that our internal control over financial reporting is effective as required by Section 404. This could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statements.

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

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

- a classified board of directors with staggered three-year terms;
- directors only to be removed for cause;
- an affirmative vote of $66^{2}/_{3}\%$ 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 jurisdictions by reason of our activities and operations, and it is also possible that taxing authorities in any such jurisdictions could assert that we are subject to greater taxation than we currently anticipate. Any such non-Bermudan tax liability could materially adversely affect our results of operations.

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

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

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

assess interest and penalties, it would increase our consolidated tax liability, which could adversely affect our financial condition, results of operations and cash flows.

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

In late 2017, the EU Economic and Financial Affairs Council, or ECOFIN, released a list of non-cooperative jurisdictions for tax purposes. The stated aim of this list, and accompanying report, was to promote the EU's view for good governance worldwide in order to maximize efforts to prevent tax fraud and tax evasion. While Bermuda was not on the original EU list of non-cooperative jurisdictions, it committed to address EU concerns relating to economic substance by December 31, 2018. In accordance with that commitment, Bermuda enacted the Economic Substance Act 2018, or the Substance Act, requiring certain entities in Bermuda engaged in "relevant activities" to maintain a substantial economic presence in Bermuda and to satisfy economic substance requirements commencing as of July 1, 2019. The list of "relevant activities" includes carrying on as a business in any one or more of: banking, insurance, fund management, financing, leasing, headquarters, shipping, distribution and service center, intellectual property and holding entities. Under the Substance Act, any entity that must satisfy economic substance requirements but fails to do so could face automatic disclosure to competent authorities in the EU of the information filed by the entity with the Bermuda Registrar of Companies in connection with the economic substance requirements and may also face financial penalties, restriction or regulation of its business activities or may be struck as a registered entity in Bermuda. Bermuda has not yet provided guidance clarifying its "relevant activities" requirements under the Substance Act. Accordingly, we cannot predict the specifics of Bermuda's current or future economic substance requirements on our business, which may impact the manner and jurisdictions in which we operate, which could adversely affect our business, financial condition or results of operations.

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, 2019, which could result in adverse U.S. federal income tax consequences to U.S. investors in our common shares.

Because we did not to earn revenue from our business operations during the year ended December 31, 2018 and do not expect to do so for the year ended December 31, 2019, and because our sole source of income had been and currently is interest on bank accounts held by us, we believe we will likely be classified as a "passive foreign investment company," or PFIC, for the taxable year ended December 31, 2019. 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 common shares, we will continue to be treated as a PFIC with respect to such U.S. Holder in all succeeding years during which the U.S. Holder owns the common shares, regardless of whether we continue to meet the PFIC test described above, unless the U.S. Holder makes or has made a specified election and we cease to be a PFIC. 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 (or another entity taxable as a corporation) created or organized under the laws of the United States, any state thereof, or the District of Columbia;
- an estate, the income of which is subject to U.S. federal income tax regardless of its source; or
- a trust that (i) is subject to the supervision of a U.S. court and the control of one or more "United States persons" (within the meaning of Section 7701(a)(30) of the 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 common shares, certain adverse U.S. federal income tax consequences could apply to such U.S. Holder, including (i) the treatment of all or a portion of any gain on disposition as ordinary income, (ii) the application of a deferred interest charge on such gain and the receipt of certain dividends and (iii) the obligation to comply with certain reporting requirements.

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

We believe we will likely be classified as a controlled foreign corporation for the taxable year ended December 31, 2019. Even if we were not classified as a controlled foreign corporation, if our group includes one or more U.S. subsidiaries, certain of our non-U.S. subsidiaries could be treated as controlled foreign corporations. If a U.S. Holder is treated as owning (directly, indirectly or constructively) at least 10% of the value or voting power of our common shares, such U.S. Holder may be treated as a "United States shareholder" with respect to us (if we are classified as a controlled foreign corporation) and each controlled foreign corporation in our group (if any). A United States shareholder of a controlled foreign corporation may be required to annually report and include in its U.S. taxable income its pro rata share of "Subpart F income," "global intangible low-taxed income," or GILTI, and investments in U.S. property by controlled foreign corporations, regardless of whether we make any distributions. An individual that is a United States shareholder with respect to a controlled foreign corporation generally would not be allowed certain tax deductions or foreign tax credits that would be allowed to a United States shareholder that is a U.S. corporation. Failure to comply with these reporting obligations may subject 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 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 common shares.

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

In 2017, the U.S. government enacted comprehensive tax legislation that includes significant changes to the taxation of business entities, or the Tax Reform Act. These changes include, among others, a permanent reduction to the corporate income tax rate, introduction of the GILTI provision, limiting interest deductions, adopting elements of a territorial tax system and introducing certain anti-base erosion provisions. We continue to examine the impact this tax reform legislation may have on our business. The effect of the Tax Reform Act on our business, whether adverse or favorable, is uncertain, and may not become evident for some period of time. U.S. Holders should consult with their legal and tax advisors regarding any such legislation and the potential tax consequences of investing in our common shares.

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

Issuance of Unregistered Equity Securities

On June 6, 2019, we issued an aggregate of 94,284 Class A common shares to the former shareholders of Primatope, having an aggregate value of approximately $1.5 million, as payment, in part, for the achievement of the final milestone after the closing of the acquisition, 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 of the IPO and on June 22, 2018, we issued and sold an additional 1,006,425 Class A common shares pursuant to the exercise in part 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 the 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 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	4/27/18	
3.2	Amended and Restated Bye-Laws of Kiniksa Pharmaceuticals, Ltd.	8-K	001-38492	3.1	5/29/18	
4.1	Specimen Share Certificate evidencing the Class A common shares	S-1/A	333-224488	4.1	5/14/18	
4.2	Second Amended and Restated Investors' Rights Agreement, dated as of February 9, 2018	S-1	333-224488	3.1	4/27/18	
31.1	Rule 13a-14(a) / 15d-14(a) Certification of Chief Executive Officer					*
31.2	Rule 13a-14(a) / 15d-14(a) Certification of Chief Financial Officer					*
32.1	Section 1350 Certification of Chief Executive Officer					**
32.2	Section 1350 Certification of Chief Financial Officer					**
101.INS	XBRL Instance Document					***
101.SCH	XBRL Taxonomy Extension Schema Document					***
101.CAL	XBRL Taxonomy Extension Calculation Linkbase Document					***
101.DEF	XBRL Extension Definition Linkbase Document					***
101.LAB	XBRL Taxonomy Label Linkbase Document					***
101.PRE	XBRL Taxonomy Extension Presentation Linkbase Document					***

* Filed herewith
** Furnished herewith
*** Submitted electronically herewith

SIGNATURES

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

KINIKSA PHARMACEUTICALS, LTD.

Date: August 13, 2019

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(s) 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(s) 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.

August 13, 2019

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

Exhibit 31.2

CERTIFICATIONS

I, Chris Heberlig, 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(s) 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(s) 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.

August 13, 2019

/s/ Chris Heberlig
Chris Heberlig
Chief 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 June 30, 2019 (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.

August 13, 2019

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

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, Chris Heberlig, Chief Financial Officer of Kiniksa Pharmaceuticals, Ltd. (the "Company"), hereby certify, pursuant to 18 U.S.C. §1350, as adopted pursuant to §906 of the Sarbanes-Oxley Act of 2002, that, to the best of my knowledge:

(1) The Quarterly Report on Form 10-Q of the Company for the period ended June 30, 2019 (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.

August 13, 2019

/s/ Chris Heberlig
Chris Heberlig
Chief Financial Officer