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Filed Pursuant to Rule 424(b)(4)
Registration No. 333-229394

Prospectus

2,654,984 shares

Class A common shares

This is a public offering of our Class A common shares. All 2,654,984 Class A common shares are being sold by us. The public offering price per share is $18.26.

Our Class A common shares are listed on The Nasdaq Global Select Market under the symbol "KNSA." The last reported sale price of our Class A common shares on January 30, 2019 was $18.26 per share.

We are an "emerging growth company" as defined in Section 2(a) of the Securities Act of 1933, as amended, or the Securities Act, as modified by the Jumpstart Our Business Startups Act of 2012, and a smaller reporting company, as defined in the rules promulgated under the Securities Act, and as such have elected to comply with certain reduced public company reporting requirements for this prospectus and our other filings with the Securities and Exchange Commission. See "Prospectus summary—Implications of being an emerging growth company and a smaller reporting company."

Investing in our Class A common shares involves risk. See "Risk factors" beginning on page 12 to read about factors you should consider before buying our Class A common shares.

Neither the Securities and Exchange Commission nor any other regulatory body has approved or disapproved of these securities or passed upon the accuracy or adequacy of this prospectus. Any representation to the contrary is a criminal offense.

	Per share	Total
Offering price	$ 18.26	$ 48,480,008
Underwriting discounts and commissions(1)	$ 1.00	$ 2,666,400
Proceeds, before expenses, to Kiniksa Pharmaceuticals, Ltd.	$ 17.26	$ 45,813,607

(1) See "Underwriting" beginning on page 206 for additional information regarding underwriting compensation.

We have four classes of common shares outstanding: Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares. All classes of our common shares are economically equivalent to each other. The rights of the holders of our Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares are identical, except with respect to voting, conversion and transferability. Each Class A common share is entitled to one vote and is not convertible into any other class of our share capital. Each Class B common share is entitled to ten votes and is convertible at any time at the election of the holder into one Class A common share or one Class B1 common share and automatically converts into a Class A common share upon transfer to an unaffiliated party. The rights of the holders of our Class A1 common shares and Class B1 common shares are identical, except with respect to conversion and transferability. Each Class A1 common share and Class B1 common share has no associated voting rights. Each Class A1 common share is convertible into one Class A common share, subject to certain limitations, as described in this prospectus. Each Class B1 common share is convertible into one Class A common share or one Class B common share, subject to certain limitations, as described in this prospectus and automatically converts into a Class A common share upon transfer to an unaffiliated party. Immediately following this offering and the concurrent private placement described in this prospectus, the holders of our Class A common shares will account for 28.5% of our aggregate voting power and the holders of our Class B common shares will account for the remaining 71.5% of our aggregate voting power. See "Description of share capital—Common shares" for more information on the rights of the holders of our Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares.

One or more entities managed by Baker Bros. Advisors LP, an affiliate of certain of our directors, have agreed to purchase 2,000,000 Class A1 common shares in a private placement exempt from the registration requirements of the Securities Act at a sale price equal to the public offering price of our Class A common shares in this offering, or the concurrent private placement. The underwriters will serve as placement agents for the concurrent private placement and receive a placement agent fee equal to a percentage of the total purchase price of the private placement shares, which percentage will be equal to the percentage discount the underwriters will receive on shares sold in this public offering. The consummation of the concurrent private placement is contingent on the closing of this offering and the satisfaction of certain other customary conditions. However, the consummation of this offering is not contingent on the consummation of the concurrent private placement.

We have granted the underwriters the option to purchase up to an additional 398,247 of our Class A common shares for a period of 30 days after the date of this prospectus.

The underwriters expect to deliver the Class A common shares to investors against payment on or about February 4, 2019.

J.P. Morgan **Goldman Sachs & Co. LLC** **Barclays**
Wedbush PacGrow **JMP Securities**

Prospectus dated January 31, 2019

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We have not, and the underwriters have not, authorized anyone to provide any information or to make any representations other than those contained in this prospectus or in any free writing prospectus prepared by or on behalf of us or to which we have referred you. We take no responsibility for, and can provide no assurance as to the reliability of, any other information that others may give you. This prospectus is an offer to sell only the shares offered hereby, and only under circumstances and in jurisdictions where it is lawful to do so. The information contained in this prospectus or in any applicable free writing prospectus is current only as of its date, regardless of its time of delivery or any sale of our Class A common shares. Our business, financial condition, results of operations and prospects may have changed since that date.

For investors outside the United States: We have not, and the underwriters have not, done anything that would permit this offering or possession or distribution of this prospectus in any jurisdiction where action for that purpose is required, other than in the United States. Persons outside the United States who come into possession of this prospectus must inform themselves about, and observe any restrictions relating to,

the offering of the Class A common shares and the distribution of this prospectus outside the United States.

Trademarks

We own or have rights to trademarks that we use in connection with the operation of our business, including Kiniksa® and ARCALYST®. Kiniksa® is a trademark of Kiniksa Pharmaceuticals, Ltd. and ARCALYST® is a trademark of Regeneron Pharmaceuticals, Inc. Solely for convenience, trademarks, service marks and trade names referred to in this prospectus, including Kiniksa and ARCALYST, are listed without the ®, SM and ™ symbols. We will assert, to the fullest extent under applicable law, our rights to our intellectual property. Trademarks, service marks and trade names of third parties are the intellectual property of such parties.

Prospectus summary

This summary highlights information contained elsewhere in this prospectus. This summary does not contain all of the information you should consider before investing in our Class A common shares. You should read this entire prospectus carefully, especially the sections entitled "Risk factors" and "Management's discussion and analysis of financial condition and results of operations" and our consolidated financial statements and the related notes appearing at the end of this prospectus, before making an investment decision. This prospectus includes forward-looking statements that involve risks and uncertainties. See "Special note regarding forward-looking statements."

As used in this prospectus, unless the context otherwise requires, references to "we," "us," "our," the "Company" and "Kiniksa" refer to Kiniksa Pharmaceuticals, Ltd. and its consolidated subsidiary, together.

Overview

We are a 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 five product candidates, across various stages of development, focused on autoinflammatory and autoimmune conditions. We have three clinical-stage product candidates and two pre-clinical-stage product candidates. We follow a disciplined and methodical approach to selectively identify, discover and acquire product candidates with strong biologic rationales or validated mechanisms of action. We believe that each of our product candidates has the potential to address multiple indications.

Our portfolio of product candidates offers multiple development opportunities. By modulating different parts of the innate and adaptive immune system, we believe these product candidates together have the potential to provide a variety of mechanisms to address multiple devastating diseases.

Our programs

- ***Rilonacept*** is a protein cytokine trap for inhibiting interleukin-1a, or IL-1a, and interleukin-1b, or IL-1b. Cytokines are small proteins that play a key role in cell signaling. Rilonacept is approved by the U.S. Food and Drug Administration, or the FDA, for the treatment of cryopyrin-associated periodic syndromes, or CAPS, and has been commercially sold as ARCALYST by Regeneron Pharmaceuticals, Inc., or Regeneron, for this indication since 2008. We licensed rilonacept in 2017 from Regeneron. We are initially developing rilonacept for the treatment of recurrent pericarditis, a debilitating inflammatory cardiovascular disease. We are not aware of any therapy currently approved by the FDA for the treatment of recurrent pericarditis. We are enrolling a single, pivotal, global, Phase 3 clinical trial in recurrent pericarditis, named RHAPSODY. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data in the second half of 2020. RHAPSODY is a double-blind, placebo-controlled, randomized-withdrawal, or RW, design study with open-label extension which is designed to assess the efficacy and safety of rilonacept treatment in subjects with recurrent pericarditis. We also have an ongoing open-label Phase 2 proof-of-concept clinical trial in recurrent pericarditis, for which we recently completed enrollment. In December 2018, we reported interim data from the Phase 2 trial. As of the November 1, 2018 data cutoff date, interim data from 12 symptomatic subjects participating in one portion of the Phase 2 trial showed a reduction in both c-reactive protein, an inflammation biomarker, and reported pain. As of the cutoff date, 10 of the subjects had completed the 6-week base treatment period and entered into the optional 18-week extension period. Four of the 10 subjects had completed the optional 18-week extension period. All subjects showed a persistent clinical response as measured by c-reactive protein and pain levels at each measurement point during the study. Rilonacept

has been generally well-tolerated in the trial, with adverse events, or AEs, consistent with the FDA-approved label for the treatment of CAPS. The most common AEs were gastrointestinal disorders and injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment. Infections are reported in the rilonacept label. We expect to present additional data from this trial at the American College of Cardiology 68th Annual Scientific Session & EXPO 2019, or ACC, in the first half of 2019.

- ***Mavrilimumab*** is a monoclonal antibody that antagonizes the signaling of granulocyte macrophage colony stimulating factor, or GM-CSF. We are focusing our initial development efforts for mavrilimumab on giant cell arteritis, or GCA, an inflammatory disease of the blood vessels with unmet medical need that can lead to blindness if left untreated. MedImmune Limited, or MedImmune, initially developed mavrilimumab for the treatment of rheumatoid arthritis, or RA. MedImmune's Investigational New Drug application, or IND, for the clinical development of mavrilimumab for the treatment of RA was initially put on clinical hold in 2010 before human data had been generated due to certain effects that were observed in non-clinical studies, which coincides with a theoretical risk of developing pulmonary alveolar proteinosis, or PAP, possibly in the setting of GM-CSF inhibition. Since then, in 2014, the FDA acknowledged that clinical studies in refractory RA may be appropriate based on MedImmune's clinical studies in Europe in which it dosed over 550 RA patients with mavrilimumab with no evidence of PAP. MedImmune has since withdrawn the IND for mavrilimumab for the treatment of RA, and we submitted a new IND with the FDA for the study of mavrilimumab in GCA. The FDA initially placed our IND on clinical hold due to its request for additional information regarding the 510(k)-cleared delivery device to be used in our Phase 2 clinical trial. We have since provided the FDA with the requested information and our IND is now active. We plan for U.S. subjects to be included in our ongoing, double-blind, randomized, placebo-controlled, global Phase 2 proof-of-concept clinical trial, for which we have commenced dosing in multiple countries. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data 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 OSMRb. We plan to study KPL-716 in a variety of pruritic, inflammatory, and fibrotic indications driven by these cytokines, and we believe KPL-716 is the only monoclonal antibody in development that simultaneously targets both pathways. At the European Association of Dermatology and Venereology congress in September 2018, we presented results from the randomized, double-blind, placebo-controlled, single-ascending-dose, sequential-group portion of the Phase 1a/1b clinical trial in healthy volunteers and in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. In this trial, single intravenous, or IV, and subcutaneous, or SC, doses were well-tolerated. The results provided an early signal in efficacy in reducing pruritus (assessed by the Worst-Itch Numerical Rating Scale), as well as reducing inflammation and disease severity (assessed by Eczema Area Severity Index, or EASI) in atopic dermatitis subjects after a single dose of KPL-716 in a placebo-controlled, exploratory efficacy assessment (20 subjects were randomized 1:1 and received either 7.5 mg/kg IV of KPL-716 or placebo). To help us to understand whether KPL-716 could be a competitive therapeutic in atopic dermatitis, if approved, we are enrolling a 12-week, repeated single-dose cohort as an additional part of the Phase 1b portion of the Phase 1a/1b clinical trial in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. This cohort is designed to evaluate safety, tolerability, pharmacokinetics and immunogenicity, and it will allow us to conduct an exploratory efficacy analysis on both pruritus as well as disease severity response markers (assessed by EASI). We expect to report top-line data from this cohort in the second half of 2019. We also believe that the single-dose results from the Phase 1a/1b study provide proof-of-principle for KPL-716's potential

to treat a spectrum of diseases that may have pruritus mediated by IL-31 and support our plans to advance KPL-716 into multiple diseases where chronic pruritus plays a role in patient symptomatology. We plan to initiate an adaptive design Phase 2a/2b clinical trial in prurigo nodularis in the first half of 2019 and expect to report top-line data from the first part of this trial in the first half of 2020. We also plan to initiate an exploratory, pilot Phase 2 clinical trial in the first half of 2019 designed to explore the role of IL-31 and OSM in a number of diseases characterized by chronic pruritus and to report top-line data from this trial in the second half of 2020.

- ***KPL-045*** is a monoclonal antibody inhibitor of the CD30/CD30L interaction, a T-cell co-stimulatory receptor involved in activated T-memory cell function. We are continuing our pre-clinical activities in KPL-045 in inflammatory diseases driven by T-cell-dependent autoantibody generation and dysregulated T_H effector memory responses and expect to file an IND with the FDA in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020.

- ***KPL-404*** is a monoclonal antibody inhibitor of the CD40/CD40L interaction, a central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity. We are continuing our pre-clinical activities in KPL-404 in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020. In January 2019, we exercised our exclusive option to acquire all of the outstanding capital stock of Primatope Therapeutics, Inc., or Primatope, the company that owns or controls the intellectual property related to KPL-404. We expect to close this transaction within 60 days of the option exercise subject to a supplemental due diligence period. If we do not close this transaction, our license to the intellectual property controlled by Primatope to research, develop and manufacture KPL-404 will terminate and we will cease the development of KPL-404.

The following table summarizes our current pipeline of product candidates:

Program & Target	Indication	Phase (Preclin / 1 / 2 / 3)	Status	Rights
Rilonacept[1] IL-1α & IL-1β	Recurrent Pericarditis (RP)		• Enrolling single, pivotal Phase 3 trial in recurrent pericarditis; top-line data expected in 2H 2020 • Phase 2 trial data presentation at ACC expected in 1H 2019	Worldwide (excluding MENA)
Mavrilimumab GM-CSFRα	Giant Cell Arteritis (GCA)		• Enrolling global Phase 2 proof-of-concept trial • Top-line data expected in 2H 2020	Worldwide
KPL-716 OSMRβ	Prurigo Nodularis (PN)		• Plan to initiate adaptive design Phase 2a/2b trial in PN in 1H 2019 • Top-line data from Phase 2a expected in 1H 2020	Worldwide
	Chronic Idiopathic Urticaria, Chronic Idiopathic Pruritus, Lichen Planus, Lichen Simplex Chronicus & Plaque Psoriasis		• Plan to initiate Phase 2 exploratory pilot trial in multiple diseases characterized by chronic pruritus in 1H 2019 • Top-line data from Phase 2 trial expected in 2H 2020	
	Atopic Dermatitis (AtD)		• Top-line data from repeated-single-dose Phase 1b trial expected in 2H 2019	
KPL-045[2] CD30L	Autoimmune		• IND filing planned for 2H 2019 • Plan to initiate Phase 1 trial in 1H 2020	Worldwide
KPL-404[2 3] CD40	Autoimmune		• IND filing planned for 2H 2019 • Plan to initiate Phase 1 trial in 1H 2020	Worldwide

(1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication.

(2) We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.

(3) Subject to closing the acquisition of Primatope.

In addition to the indications described above, we plan to evaluate rilonacept, mavrilimumab and KPL-716 in other indications. We plan to be opportunistic in our business development activities to identify and potentially acquire the rights to additional programs or companies that could expand our existing portfolio and/or platform capability. We have also initiated our own internal research efforts to discover and develop molecules to address areas of unmet medical need.

We currently plan to directly commercialize our product candidates, if approved, in the United States and select international markets. In parallel with our product development timelines, we plan to build our own commercial and operational organizations around the world as appropriate. We anticipate building targeted medical affairs and sales teams focused on specialist physicians who treat the patient populations addressed by our product candidates.

Our team

We have assembled an experienced management team with a successful track record. Our team has expertise across the spectrum of global drug discovery, development, manufacturing and commercialization activities in diseases within both large and orphan indications. Our Chairman and Chief Executive Officer, Sanj K. Patel, has more than 25 years of scientific, clinical and commercial experience in the pharmaceutical and biotechnology industries. Our Chief Medical Officer, John F. Paolini, M.D., Ph.D., has more than 17 years of experience planning, operating and executing clinical development programs across a range of disease indications from orphan diseases to large cardiovascular diseases, and ten years as a practicing cardiologist.

Our strategy

Our vision is to build a fully-integrated global biopharmaceutical company by discovering, acquiring, developing and commercializing life-changing therapies for debilitating diseases. We are developing a pipeline of novel drug product candidates for the treatment of autoinflammatory and autoimmune diseases, and we aim to be an industry leader in these areas. We are pursuing multiple programs in parallel, with the goal of delivering safe and effective therapies to patients as efficiently as possible.

Critical components of our business strategy include the following:

- **Efficiently and rapidly advance our product candidates through the development process.** We believe that our product candidates have the potential to address significant unmet medical needs and intend to develop them as efficiently and rapidly as possible.
- **Commercialize our product candidates to bring new or improved therapies to patients in need.** We intend to market and commercialize our product candidates, if approved, in the United States and select international markets by developing our own sales, marketing, medical affairs and reimbursement organizations. We anticipate creating a targeted sales organization that supports specialist physicians who treat these specific patient populations and plan to build out this organization as our product candidates approach potential regulatory approval. We believe this approach will allow us to effectively reach patients and prescribers that our product candidates target and leverage the commercial potential of our product candidates.
- **Maximize our existing portfolio opportunity by expanding use across multiple indications.** A core component of our approach to product development is identifying assets that each have the potential to treat multiple diseases. We aim to develop and commercialize our product candidates to produce meaningful impact for patients across relevant indications. Our assets are designed to specifically

modulate signaling pathways that are implicated across a spectrum of autoimmune and autoinflammatory conditions. We believe that all of our product candidates have potential in multiple indications.

- **Leverage our value-driven approach to identify, discover, acquire and develop new therapies.** We follow a disciplined and methodical approach to our review of new opportunities. We focus on research-based and comprehensive indication mapping exercises to categorize and prioritize indications of interest. We evaluate a variety of factors for potential product candidates, technologies and discovery targets, including biologic rationale for addressing the disease, potential for regulatory approval, commercial viability, intellectual property position, prospects for favorable pricing and reimbursement and the impact of competition. We also look at assets that could potentially address multiple indications. In building our current pipeline, we evaluated a large number of opportunities and negotiated agreements with parties for the assets that met our criteria and have in-licensed or acquired the rights to develop and commercialize five separate biologics. Going forward, we intend to be opportunistic in our business development activities.

- **Build our core capability in autoimmune and autoinflammatory diseases to establish a leadership position in the field.** Our current pipeline consists of protein therapeutic candidates across various stages of drug development, including a cytokine trap—rilonacept—and four monoclonal antibodies—mavrilimumab, KPL-716, KPL-045 and KPL-404. Both categories of therapeutics functionally inhibit signaling pathways that are implicated in autoinflammatory- or autoimmune-driven pathologies. We intend to leverage our internal discovery efforts and business development capabilities to complement our existing portfolio to build our core capability and establish a leadership position in the field.

Our capital structure

We have four classes of common shares: Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares. All classes of our common shares are economically equivalent to each other. The rights of the holders of our Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares are identical, except with respect to voting, conversion and transferability. Holders of our Class A common shares—the only class of common shares being sold in this offering—are entitled to one vote per Class A common share, while holders of our Class B common shares are entitled to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares have no associated voting rights. Following this offering and the concurrent private placement, the Class A common shares will account for 28.5% of our aggregate voting power and the Class B common shares will account for the remaining 71.5% of our aggregate voting power. In addition, following the completion of the concurrent private placement, we will have 14,995,954 Class A1 common shares outstanding and 16,057,618 Class B1 common shares outstanding. See "Principal shareholders" and "Description of share capital" for more information on beneficial ownership immediately following this offering and the concurrent private placement.

As a result of the Class A common shares and Class B common shares that they hold, our executive officers and certain other members of our senior management are able to exercise voting rights with respect to an aggregate of 857,911 Class A common shares and 4,244,005 Class B common shares, which will collectively represent 66.8% of the voting power of our outstanding share capital immediately following this offering and the concurrent private placement. As a result, our executive officers and certain other members of our senior management will have the ability to control the outcome of all matters submitted to our shareholders for approval, including the election, removal and replacement of directors and any merger, consolidation, or sale of all or substantially all of our assets. However, this percentage

may change depending on any conversion of our Class A1 common shares, Class B1 common shares or Class B common shares. Each holder of our Class A1 common shares may elect to convert its non-voting Class A1 common shares into voting Class A common shares at any time, unless, 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. Each holder of our Class B1 common shares may elect to convert its non-voting Class B1 common shares into voting Class A common shares or Class B common shares at any time, unless, 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. Any such holder of Class A1 common shares or Class B1 common shares has the right to increase, decrease or waive this beneficial ownership limitation at its sole discretion by providing us with 61-days' prior written notice. As a result, upon 61-days' prior written notice, entities managed by Baker Bros. Advisors LP, or Baker Brothers, could convert their Class A1 common shares and Class B1 common shares into Class A common shares and Class B common shares, respectively, which in the aggregate would result in such entities holding 73.7% of the voting power of our outstanding share capital following this offering and the concurrent private placement.

This concentrated control could delay, defer, or prevent a change of control, merger, consolidation, or sale of all or substantially all of our assets that our other shareholders support. Conversely, this concentrated control could allow our executive officers and certain other members of our senior management to consummate a transaction that our other shareholders do not support. See "Risk factors—Risks related to our common shares and this offering—*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 all matters submitted to our shareholders for approval. This concentration of ownership of our Class B common shares 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*."

Risks associated with our business

Our business is subject to numerous risks and uncertainties, including those highlighted in the section titled "Risk factors" immediately following this prospectus summary. Some of these risks are:

- we have a limited operating history, have never generated any product revenue, have incurred significant operating losses since our inception, expect to incur significant operating losses for the foreseeable future and may never achieve or maintain profitability;
- we may not be successful in our efforts to identify, discover, develop or acquire additional product candidates;
- we depend heavily on the success of our product candidates and cannot give any assurance that our product candidates will receive regulatory approval for any indication, which is necessary before they can be commercialized;
- we will need additional funding to complete the development and commercialization of our product candidates, if approved, and to acquire additional product candidates, and if we are unable to raise capital when needed, we could be forced to delay, reduce or eliminate our product development programs or future commercialization efforts;
- we had no involvement with or control over the pre-clinical and clinical development of our current product candidates prior to our acquisition of them, and we are dependent on the parties from whom we

licensed or acquired such product candidates having conducted their research and development in accordance with the applicable protocols and standards, accurately reported the results of all clinical trials conducted prior to our acquisition and correctly collected and interpreted the data from these trials;

- we have acquired product candidates with positive clinical data in diseases other than our target indications, and we cannot be certain that our product candidates will prove to be effective in treating our target indications;
- we rely, and expect to continue to rely, on third parties to conduct our clinical trials and to manufacture our product candidates for pre-clinical and clinical testing, including our sole source of supply for each of our active pharmaceutical ingredients, and those third parties may not perform satisfactorily, which could delay our product development activities;
- all of our product candidates have been licensed or acquired from other parties; if we are unable to adequately protect our product candidates, or to secure and maintain freedom to operate, others could preclude us from commercializing our product candidates or compete against us more directly;
- we face significant competition from other biotechnology and pharmaceutical companies;
- concentration of ownership of the voting power of our common shares may prevent new investors in this offering from influencing significant corporate decisions and may have an adverse effect on the price of our Class A common shares; and
- we will likely be classified as a passive foreign investment company and we believe we have been classified as a controlled foreign corporation in the current taxable year and may be classified as a passive foreign investment company or controlled foreign corporation in any future taxable year, which may result in adverse U.S. federal income tax consequences to U.S. holders of our Class A common shares.

Concurrent private placement

One or more entities managed by Baker Brothers have agreed to purchase 2,000,000 of our non-voting Class A1 common shares in a private placement exempt from the registration requirements of the Securities Act at a sale price equal to the public offering price of our Class A common shares in this offering. The consummation of the concurrent private placement is contingent on the closing of this offering and the satisfaction of certain other customary conditions. The underwriters will serve as placement agents for the concurrent private placement and receive a placement agent fee equal to a percentage of the total purchase price of the private placement shares, which percentage will be equal to the percentage discount the underwriters will receive on shares sold in this public offering. The concurrent private placement is expected to close concurrently with this offering. The consummation of this offering is not contingent on the consummation of the concurrent private placement.

Our corporate information

We are an exempted company incorporated under the laws of Bermuda in July 2015. Our registered office is located in Bermuda at Clarendon House, 2 Church Street, Hamilton HM11, Bermuda. The telephone number for our registered office is +44 808-189-6257. Our website address is www.kiniksa.com. The information contained on our website is not incorporated by reference into this prospectus, and you should not consider any information contained on, or that can be accessed through, our website as part of this prospectus or in deciding whether to purchase our Class A common shares.

Implications of being an emerging growth company and a smaller reporting company

We qualify as an "emerging growth company" as defined in Section 2(a) of the Securities Act of 1933, as amended, or the Securities Act, as modified by the Jumpstart Our Business Startups Act of 2012, or the JOBS Act, and as a "smaller reporting company" as defined in rules promulgated under the Securities Act. Emerging growth companies and smaller reporting companies may take advantage of exemptions from some of the reporting requirements that are otherwise applicable to public companies. These exemptions include:

- being permitted to present only two years of audited financial statements, in addition to any required unaudited interim financial statements, with correspondingly reduced "Management's discussion and analysis of financial condition and results of operations" disclosure in this prospectus;
- not being required to provide selected financial data or supplementary financial information;
- not being required to comply with the auditor attestation requirements in the assessment of our internal control over financial reporting;
- an exemption from compliance with the requirement of the Public Company Accounting Oversight Board regarding the communication of critical audit matters in the auditor's report on our financial statements;
- reduced disclosure obligations regarding executive compensation in our periodic reports, proxy statements and registration statements; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation, 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.

As an emerging growth company, we may take advantage of the applicable exemptions until the last day of our fiscal year following the fifth anniversary of the closing of the initial public offering of our Class A common shares, or the IPO. However, we will cease to be an emerging growth company prior to the end of such five-year period if (i) we become a "large accelerated filer" as defined in Rule 12b-2 under the Securities Exchange Act of 1934, as amended, which would occur if the market value of our voting and non-voting common equity held by non-affiliates exceeds $700 million as of the last business day of our most recently completed second fiscal quarter; (ii) our annual gross revenue exceeds $1.07 billion; or (iii) we issue more than $1.0 billion of non-convertible debt in any three-year period.

As a smaller reporting company, we may take advantage of the applicable exemptions until the fiscal year following the determination that our voting and non-voting common shares held by non-affiliates is more than $250 million measured on the last business day of our second fiscal quarter, or our annual revenues are more than $100 million during the most recently completed fiscal year and our voting and non-voting common shares held by non-affiliates is more than $700 million measured on the last business day of our second fiscal quarter.

We have elected to take advantage of certain of the reduced disclosure obligations in this prospectus and in our filings with the Securities and Exchange Commission. As a result, the information that we provide to our shareholders may be different than you might receive from other public reporting companies in which you hold equity interests.

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

The offering

Class A common shares offered by us	2,654,984 shares
Option to purchase additional Class A common shares	398,247 shares
Concurrent private placement	One or more entities managed by Baker Brothers have agreed to purchase 2,000,000 of our Class A1 common shares in a private placement at a price equal to the public offering price of our Class A common shares in this offering.
Class A common shares to be outstanding after this offering and the concurrent private placement	18,452,204 shares (18,850,451 shares if the underwriters exercise their option to purchase additional Class A common shares in full)
Class B common shares to be outstanding after this offering and the concurrent private placement	4,638,855 shares
Class A1 common shares to be outstanding after this offering and the concurrent private placement	14,995,954 shares
Class B1 common shares to be outstanding after this offering and the concurrent private placement	16,057,618 shares
Total common shares to be outstanding after this offering and the concurrent private placement	54,144,631 shares (54,542,878 shares if the underwriters exercise their option to purchase additional Class A common shares in full)
Voting rights	We have four classes of common shares outstanding: Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares. Each Class A common share entitles its holder to one vote per Class A common share. Each Class B common share entitles its holder to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares do not have voting rights. Immediately following this offering and the concurrent private placement, the holders of our Class A common shares will account for 28.5% of our aggregate voting power and the holders of our Class B common shares will account for the remaining 71.5% of our aggregate voting power. See "Principal shareholders" and "Description of share capital" for additional information.

Use of proceeds	We estimate that the net proceeds from this offering and the concurrent private placement will be approximately $79.3 million (or approximately $86.2 million if the underwriters exercise their option to purchase additional Class A common shares in full), based on the offering price of $18.26 per share. We intend to use the net proceeds from this offering and the concurrent private placement to advance the clinical development of rilonacept, mavrilimumab and KPL-716, to fund other research and development activities and for working capital and general corporate purposes. See "Use of proceeds" beginning on page 86.
Risk factors	See "Risk factors" beginning on page 12 and the other information included in this prospectus for a discussion of factors you should consider carefully before deciding to invest in our Class A common shares.
The Nasdaq Global Select Market symbol	"KNSA"

The total number of common shares to be outstanding after this offering and the concurrent private placement is based on 15,797,220 Class A common shares, 4,638,855 Class B common shares, 12,995,954 Class A1 common shares and 16,057,618 Class B1 common shares outstanding as of December 31, 2018. This amount excludes:

- 5,960,939 Class A common shares issuable upon exercise of share options outstanding as of December 31, 2018, at a weighted average exercise price of $10.25 per share;
- 3,175,665 Class A common shares available for future issuance under our 2018 Incentive Award Plan, or the 2018 Plan, as of December 31, 2018, as well as common shares that become available pursuant to provisions in the 2018 Plan that automatically increase the share reserve under the 2018 Plan as described in "Executive and director compensation—Executive compensation plans—2018 incentive award plan"; and
- 648,660 Class A common shares available for future issuance under our 2018 Employee Share Purchase Plan, or the ESPP, as of December 31, 2018, as well as common shares that become available pursuant to provisions in the ESPP that automatically increase the share reserve under the ESPP as described in "Executive and director compensation—Executive compensation plans—2018 employee share purchase plan".

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

- no exercise of outstanding share options after December 31, 2018; and
- no exercise by the underwriters of their option to purchase additional Class A common shares.

Summary consolidated financial data

You should read the following summary consolidated financial data together with our consolidated financial statements and the related notes appearing at the end of this prospectus and the "Selected consolidated financial data" and "Management's discussion and analysis of financial condition and results of operations" sections of this prospectus. We have derived the consolidated statement of operations data for the years ended December 31, 2016 and 2017 from our audited consolidated financial statements appearing at the end of this prospectus. We have derived the consolidated statement of operations data for the nine months ended September 30, 2017 and 2018 and the consolidated balance sheet data as of September 30, 2018 from our unaudited consolidated financial statements appearing at the end of this prospectus, which have been prepared on the same basis as the audited consolidated financial statements. In the opinion of management, the unaudited data reflects all adjustments, consisting only of normal recurring adjustments, necessary for a fair statement of the financial information in those statements. Our historical results are not necessarily indicative of results that may be expected in any future period, and our results for any interim period are not necessarily indicative of results that may be expected for any full year.

	Year ended December 31,		Nine months ended September 30,	
	2016	2017	2017	2018
	(in thousands, except share and per share data)			
Consolidated Statement of Operations Data:				
Operating expenses:				
Research and development	$ 17,439	$ 56,357	$ 26,426	$ 50,475
General and administrative	6,563	9,043	6,263	13,550
Total operating expenses	24,002	65,400	32,689	64,025
Loss from operations	(24,002)	(65,400)	(32,689)	(64,025)
Interest income	65	529	396	2,992
Loss before benefit (provision) for income taxes	(23,937)	(64,871)	(32,293)	(61,033)
Benefit (provision) for income taxes	(36)	(2)	121	386
Net loss	$ (23,973)	$ (64,873)	$ (32,172)	$ (60,647)
Net loss per share attributable to common shareholders—basic and diluted(1)	$ (91.61)	$ (35.85)	$ (19.21)	$ (2.62)
Weighted average common shares outstanding—basic and diluted(1)	261,695	1,809,751	1,675,133	23,174,841

(1) See Note 11 to our annual consolidated financial statements and Note 10 to our interim consolidated financial statements included elsewhere in this prospectus for further details on the calculation of basic and diluted net loss per share attributable to common shareholders.

	As of September 30, 2018	
	Actual	As adjusted(2)
	(in thousands)	
Consolidated Balance Sheet Data:		
Cash, cash equivalents and short-term investments	$ 337,863	$ 417,188
Working capital(1)	313,507	392,832
Total assets	347,101	426,426
Total shareholders' equity	318,913	398,238

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

(2) The as adjusted balance sheet data give effect to our issuance and sale of 2,654,984 Class A common shares in this offering and our issuance and sale of 2,000,000 Class A1 common shares in the concurrent private placement, each at the offering price of $18.26 per share, after deducting the underwriting discounts and commissions, placement agent fees and estimated offering expenses payable by us.

Risk factors

Investing in our Class A common shares involves a high degree of risk. You should carefully consider the risks described below, as well as the other information in this prospectus, including our consolidated financial statements and the related notes and "Management's discussion and analysis of financial condition and results of operations," before deciding whether to invest in our Class A common shares. The occurrence of any of the events or developments described below could adversely affect our business, financial condition, results of operations and growth prospects. In such an event, the market price of our Class A common shares could decline, and you may lose all or part of your investment. Additional risks and uncertainties not presently known to us or that we currently deem immaterial also may impair our business operations.

Risks related to our financial position and capital needs

We are a 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 any Phase 3 or other pivotal clinical trials, obtain regulatory approvals, manufacture a commercial-scale drug, or conduct sales and marketing activities. We currently generate no revenue from sales of any products, and we may never be able to develop or commercialize a marketable product. Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk. Typically, it takes many years to develop one new drug from the time it is discovered to when it is available for treating patients, and development may cease for a number of reasons. Consequently, predictions about our future success or viability could be more accurate if we had a longer operating history.

We have incurred significant losses related to expenses for research and development and our ongoing operations. Our net losses for the years ended December 31, 2016 and 2017 and the nine months ended September 30, 2017 and 2018 were $24.0 million, $64.9 million, $32.2 million and $60.6 million, respectively. As of September 30, 2018, we had an accumulated deficit of $151.6 million. We expect to continue to incur losses for the foreseeable future, and we anticipate these losses will increase substantially as we:

- continue our research and pre-clinical and/or clinical development of our product candidates, including our single, pivotal, global, Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, named RHAPSODY, our ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept in recurrent pericarditis, our global Phase 2 proof-of-concept clinical trial with mavrilimumab for the treatment of GCA and the repeated single-dose cohort portion of our ongoing Phase 1b clinical trial of KPL-716 in subjects with atopic dermatitis;

- advance the development of our programs, including our plans for advancing KPL-716 into multiple chronic pruritic diseases, with an adaptive design Phase 2a/2b clinical trial with KPL-716 in prurigo

nodularis as well as a Phase 2 exploratory pilot study in five other diseases characterized by chronic pruritus;

- initiate additional pre-clinical studies and clinical trials for our product candidates;
- increase our manufacturing needs or add additional manufacturers or suppliers;
- seek regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;
- establish a sales, marketing and distribution infrastructure to commercialize any products for which we may obtain marketing approval;
- seek to identify, assess and study new or expanded indications for our product candidates, new or alternative dosing levels and frequency for our product candidates, and/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, including the payment of up to $18 million in a combination of cash and our Class A common shares if we close the acquisition of all of the outstanding capital stock of Primatope;
- seek to maintain, protect and expand our intellectual property portfolio;
- seek to attract and retain skilled personnel;
- create additional infrastructure to support our operations as a public company and our product development and planned future commercialization efforts; and
- experience any delays or encounter issues with any of the above, including but not limited to failed trials, complex results, safety issues, other regulatory challenges that require longer follow-up of existing trials, additional major trials or additional supportive trials in order to pursue marketing approval.

Further, the net losses we incur may fluctuate significantly from quarter to quarter and year to year, such that a period-to-period comparison of our results of operations may not be a good indication of our future performance. 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 pre-clinical and clinical development, including our multiple ongoing and planned 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, time and cost necessary for completing our single, pivotal, global, Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, named RHAPSODY, our ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept in recurrent pericarditis, our global Phase 2 proof-of-concept clinical trial with mavrilimumab for the treatment of GCA, and the repeated single-dose cohort portion of our ongoing Phase 1b clinical trial of KPL-716 in subjects with atopic dermatitis, and our plans for advancing KPL-716 into multiple chronic pruritic diseases, with an adaptive design Phase 2a/2b clinical trial with KPL-716 in prurigo nodularis as well as a Phase 2 exploratory pilot study in five other diseases characterized by chronic pruritus;

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

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

- the costs and timing of transferring manufacturing technology to third-party manufacturers, producing product candidates to support clinical trials and preparing to manufacture mavrilimumab and KPL-716 on a commercial scale, as well as producing rilonacept in potential new final form configurations;

- the timing and amount of milestone and other payments we must make under our agreements with Regeneron, MedImmune, Biogen MA Inc., or Biogen, Novo Nordisk A/S, or Novo Nordisk, and the other third parties from whom we have acquired or in-licensed our product candidates or from whom we may in the future acquire or in-license product candidates or if we close the acquisition of all of the outstanding capital stock of Primatope;

- our ability to successfully commercialize any of our product candidates, including the cost and timing of forming and expanding our sales organization and marketing capabilities;

- the amount of sales revenues from our product candidates, if approved, including the sales price and the availability of coverage and adequate third-party reimbursement;

- competitive and potentially competitive products and technologies and patients' receptivity to our product candidates and the technology underlying them in light of competitive products and technologies;
- the cash requirements of any future acquisitions, developments or discovery of additional product candidates, including any licensing, 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 and/or frequency for our product candidates, and/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 products;
- the costs associated with being a public company;
- our need and ability to hire additional personnel; and
- the receptivity of the capital markets to financings by biotechnology companies generally and companies with product candidates and technologies such as ours specifically.

Any additional fundraising efforts may divert our management from their day-to-day activities, which may adversely affect our ability to develop and commercialize our product candidates. 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 pre-clinical 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, including purchasers in this offering, restrict our operations or require us to relinquish rights to our technologies or product candidates.

Until such time as we can generate substantial product revenues, if ever, we expect to finance our cash needs through securities offerings or debt financings, or possibly, license and collaboration agreements or research grants. The terms of any financing may adversely affect the holdings or the rights of our shareholders and our issuance of additional securities, whether equity or debt, or the possibility of such issuance, may cause the market price of our Class A common shares to decline. The sale of additional equity or convertible securities would dilute all of our shareholders, including your ownership interest. The incurrence of indebtedness would result in increased fixed payment obligations and we may be required to agree to certain restrictive covenants, such as limitations on our ability to incur additional debt, limitations on our ability to acquire, sell or license intellectual property rights and other operating restrictions that could adversely impact our ability to conduct our business. We could also be required to seek funds through arrangements with collaborators or otherwise at an earlier stage than otherwise would be desirable and we may be required to relinquish rights to some of our technologies, product candidates or

future revenue streams, or otherwise agree to terms unfavorable to us, any of which may have a material adverse effect on our business, operating results and prospects. If we raise funds through research grants, we may be subject to certain requirements, which may limit our ability to use the funds or require us to share information from our research and development. Raising additional capital through any of these or other means could adversely affect our business and the holdings or rights of our shareholders, and may cause the market price of our 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. Each of our product candidates require additional clinical development, management of pre-clinical, clinical or manufacturing activities, regulatory approval, obtaining adequate manufacturing supply, building of a commercial organization, and significant marketing efforts before we generate any revenue from product sales.

We have three product candidates in various stages of clinical development and two at the pre-clinical development stage. We may not be able to demonstrate that they are safe or effective in the indications for which we are studying them and they may not be approved. Although rilonacept is approved and marketed for human use for the treatment of CAPS, in the United States by Regeneron, we are studying rilonacept for the treatment of a different indication called recurrent pericarditis, which is currently in an ongoing Phase 2 proof-of-concept clinical trial for recurrent pericarditis, and we are actively recruiting and screening subjects for a single, pivotal, global, Phase 3 clinical trial for rilonacept for the treatment of recurrent pericarditis, named RHAPSODY. Mavrilimumab has been through Phase 2 clinical trials conducted by MedImmune for the treatment of RA, but our global Phase 2 proof-of-concept clinical trial with mavrilimumab is for the treatment of GCA. Our third clinical-stage product candidate, KPL-716, is currently in the repeated-single-dose cohort portion of our ongoing Phase 1b clinical trial in subjects with atopic dermatitis, and we plan to advance KPL-716 into multiple chronic pruritic diseases, with an adaptive design Phase 2a/2b clinical trial with KPL-716 in prurigo nodularis as well as a Phase 2 exploratory pilot study in five other diseases characterized by chronic pruritus, subject to clearance by applicable Institutional Review Boards, or IRBs, and/or other applicable regulatory authorities. We also have pre-clinical product candidates that will 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 indirect data primarily collected by other companies and, as applicable, our pre-clinical and/or 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 they are successful in clinical trials. If we do not receive regulatory approvals for one or more of our product candidates, we may not be able to continue our operations.

Each of our product candidates will require additional pre-clinical and/or clinical development, regulatory approval in one or more jurisdictions, obtaining manufacturing supply, capacity and expertise, building of a commercial organization, substantial investment and significant marketing efforts before we are able to generate any revenue from product sales. The success of our product candidates will depend on several factors, including the following:

- successful completion of pre-clinical studies, including toxicology studies, biodistribution studies and minimally efficacious dose studies in animals, conducted, where applicable, under the FDA's Good Laboratory Practice, or GLP, regulations;
- submission to the FDA of INDs and of clinical trial applications to foreign governmental authorities, for our product candidates to commence planned clinical trials or future clinical trials;
- successful enrollment in, and completion of, clinical trials, the design and implementation of which are agreed to by the applicable regulatory authorities, and the 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;
- sufficient supply of our product candidates from our CMOs;
- 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.

We plan to seek regulatory approval to commercialize our product candidates in the United States and potentially in foreign countries. While the scope of regulatory approval is similar in many countries, to obtain separate regulatory approval in multiple countries will require us to comply with numerous and varying regulatory requirements of each such country or jurisdiction regarding safety and efficacy and governing, among other things, clinical trials and commercial sales, pricing and distribution, and we cannot predict success in any such jurisdictions.

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

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

We are enrolling a single, pivotal, placebo-controlled, RW design, global Phase 3 clinical trial of rilonacept in subjects with symptomatic recurrent pericarditis, continuing our ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept for the treatment of recurrent pericarditis, and the repeated single-dose cohort portion of our ongoing Phase 1b clinical trial of KPL-716 in subjects with atopic dermatitis. We commenced dosing in multiple countries in a global Phase 2 proof-of-concept clinical trial with mavrilimumab for the treatment of GCA. We plan to advance KPL-716 into multiple chronic pruritic diseases, with an adaptive design Phase 2a/2b clinical trial in prurigo nodularis as well as a Phase 2 exploratory pilot study in five other diseases characterized by chronic pruritus, subject to clearance by applicable IRBs and/or other applicable regulatory authorities. We are also continuing our pre-clinical activities with KPL-045 and KPL-404 prior to initiating clinical trials. Commencing our planned clinical trials is subject to acceptance by the FDA of an IND or an IND amendment, 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.

Even after we receive and incorporate guidance from these regulatory authorities, such regulatory authorities could disagree that we have satisfied their requirements to commence our clinical trials, disagree with our interpretation of data from the relevant pre-clinical studies, clinical trials or 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 pre-clinical studies, clinical trials, CMC development, other studies or impose stricter approval conditions than we currently expect. For example, prior to us licensing mavrilimumab, MedImmune submitted an IND to the FDA to conduct a clinical trial of mavrilimumab in RA, and the FDA issued a clinical hold based on its review of certain

effects in the lungs observed in non-human primates in pre-clinical toxicity studies. However, following subsequent discussions between MedImmune and the FDA regarding the clinical hold and the availability of additional clinical safety data that MedImmune generated in human clinical trials conducted outside of the United States subsequent to the original IND submission, the FDA acknowledged that the risk/benefit assessment for investigation of mavrilimumab in a clinical trial may differ depending on the patient population studied. Specifically, the FDA acknowledged that the risk/benefit assessment for initiation of a clinical trial may be considered favorable in a patient population with high morbidity and limited effective treatment options, including refractory RA. As a result, we believed that the FDA's communications with MedImmune suggested that the FDA could find an acceptable risk/benefit for a clinical trial of mavrilimumab in the United States in GCA, a disease with high morbidity and limited treatment options. MedImmune has since withdrawn the IND for mavrilimumab for the treatment of RA. We completed our pre-IND meeting with the FDA and filed an IND for our Phase 2 proof-of-concept clinical trial of mavrilimumab in GCA. The FDA initially placed our IND on clinical hold in the United States due to its request for additional information on the 510(K)-cleared delivery device to be used in our trial. We have since provided the FDA with the requested information and our IND is now active.

Further, we could discover that our clinical trial design leads to enrollment 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. We anticipate that, to help inform the benefit-risk profile for the use of mavrilimumab in GCA, we will need to evaluate the effectiveness of different doses as well as mavrilimumab's pharmacokinetic profile. Successful completion of our clinical trials is a prerequisite to submitting a biologics license application, or BLA, to the FDA and a marketing authorization application, or MAA, to the European Medicines Agency, or EMA, for each product candidate and, consequently, to obtaining approval and initiating commercial marketing of our current and future product candidates. We do not know whether any of our clinical trials will begin or be completed on schedule, if at all.

A failure of one or more clinical trials can occur at any stage of testing, and our future clinical trials may not be successful. We may experience delays in our ongoing clinical trials and we do not know whether planned clinical trials will begin on time, will be allowed by regulatory authorities, need to be redesigned, enroll patients on time or will be completed on schedule, if at all. Events that may prevent successful or timely completion of clinical development include but are not limited to:

- inability to generate sufficient pre-clinical, toxicology or other *in vivo* or *in vitro* data to support the initiation of human clinical trials;
- delays in reaching a consensus with regulatory agencies on trial design or implementation;
- delays in establishing the appropriate dosage levels or frequency of dosing or treatment in clinical trials;
- delays in reaching, or failing to reach, agreement on acceptable terms with prospective CROs, and clinical trial sites, the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and clinical trial sites;
- difficulties in obtaining required 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 protocols amending study criteria and design;
- difficulty collaborating with patient groups and investigators;
- failure by our CROs, other third parties, or us to adhere to clinical trial requirements or to perform their obligations in a timely or compliant manner;
- failure to perform in accordance with the FDA's good clinical practices requirements, or GCPs, or applicable regulatory guidelines in other countries;
- patients not completing participation in a clinical trial or return for post-treatment follow-up;
- clinical trial sites or patients dropping out of a clinical trial;
- participating patients experiencing serious adverse events or undesirable side effects or are exposed to unacceptable health risks;
- safety issues, including occurrence of adverse events associated with the product candidate that are viewed to outweigh its potential benefits;
- difficulty in identifying the populations that we are trying to enroll in a particular trial, which may delay enrollment and reduce the power of a clinical trial to detect statistically significant results;
- changes in regulatory requirements, policies and guidance that require amending or submitting new clinical protocols;
- the cost of clinical trials of our product candidates being greater than we anticipate;
- clinical trials of our product candidates producing negative or inconclusive results, which may result in us deciding, or regulators requiring us, to conduct additional clinical trials or abandon drug development programs; and
- delays in manufacturing, testing, releasing, validating or importing/exporting sufficient stable quantities of our product candidates for use in clinical trials or the inability to do any of the foregoing.

We could encounter delays if a clinical trial is 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 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. Significant clinical trial delays could also allow our competitors to bring products to market before we do or shorten any periods during which we have the exclusive right to commercialize our product candidates and could impair our ability to commercialize our product candidates. In addition, many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates.

Clinical trials must be conducted in accordance with the laws and regulations of the FDA, European Union rules and regulations and other applicable regulatory authorities' legal requirements, regulations or guidelines, and are subject to oversight by these governmental agencies and IRBs at the medical institutions where the clinical trials are conducted. 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, European Union and countries outside of those jurisdictions, which could require that we amend trial protocols and/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 European Union and the United States may subject us to further delays and expenses as a result of increased shipment costs, additional regulatory requirements and the engagement of non-EU and non-U.S. CROs, as well as expose us to risks associated with clinical investigators who are unknown to the FDA or the EMA, and different standards of diagnosis, screening and medical care.

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

We must produce, through third parties, sufficient stable quantities of our product candidates for use in our clinical trials. Any delays in the production of our product candidates may lead to a delay in our clinical trials. If we make manufacturing or formulation changes to our product candidates or change manufacturers or manufacturing processes, 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 will need to transfer the manufacturing process of mavrilimumab to a third party to manufacture mavrilimumab for any 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 small scale manufacturing capabilities to support certain pre-clinical and early clinical development for KPL-045 and KPL-404. 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 pre-clinical and clinical development could result in additional costs to us or impair our ability to generate revenue and harm our business, financial condition and prospects significantly.

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

Identifying and qualifying patients to participate in clinical trials of our product candidates is critical to our success. The timing of our clinical trials depends in part on the speed at which we can recruit patients to participate in testing our product candidates, and we may experience delays in our clinical trials if we encounter difficulties in enrollment. Patient enrollment depends on many factors, including the size and nature of the patient population, eligibility criteria for the clinical trial, the proximity of patients to clinical sites, the design of the clinical protocol, the availability of competing clinical trials, the availability of new drugs approved for the indication the clinical trial is investigating, the risk that patients enrolled in clinical trials will drop out of the trials before completion of their treatment and clinicians' and patients' perceptions as to the safety and potential advantages of the product candidate being studied in relation to other available therapies.

Many of the conditions for which we plan to evaluate our current product candidates in the near future are in small disease populations. Accordingly, there are limited patient pools from which to draw for clinical trials.

In addition to the rarity of these diseases, the eligibility criteria of our clinical trials 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. We also may not be able to identify, recruit,

enroll and retain a sufficient number of patients to complete our clinical trials because of the perceived risks and benefits of the product candidate under trial, the proximity and availability of clinical trial sites for prospective patients and the patient referral practices of physicians. The availability and efficacy of competing therapies and clinical trials, and clinicians' and patients' perceptions as to the potential advantages of the product candidate being studied in relation to those available competing therapies and clinical trials, can also adversely impact enrollment. If patients are unwilling to participate in our trials for any reason, the timeline for recruiting patients, conducting trials and obtaining regulatory approval of potential products may be delayed, the commercial prospects of our product candidates will be harmed, and our ability to generate product revenue from any of these product candidates could be delayed or prevented. Moreover, failure to obtain and maintain patient consents can also lead to delay or prevent completion of clinical trials of our product candidates.

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

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

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

All of our product candidates modulate the immune system and carry risks associated with immunosuppression, including the 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 ongoing Phase 2 proof-of-concept clinical trial of rilonacept for recurrent pericarditis, the most common AEs were gastrointestinal disorders and injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment.

For mavrilimumab, there is a theoretical risk for the development of PAP. PAP is a rare lung disorder in which surfactant-derived lipoproteins accumulate excessively within pulmonary alveoli due to loss of GM-CSF function. The disease can range in severity from a sub-clinical reduction in diffusion capacity to significant dyspnea during mild exertion. In pre-clinical studies conducted by MedImmune, certain effects were observed in the lungs of non-human primates, which led the FDA to issue a clinical hold with respect

to MedImmune's proposed clinical trial in RA. Pre-clinical data generated to date suggest mavrilimumab does not reach the lungs in sufficient quantities to induce PAP at clinically relevant doses and human trials thus far have not shown a clinical effect on pulmonary function tests attributable to mavrilimumab. If the results of our clinical trials reveal a high or unacceptable severity and prevalence of these or other side effects, the FDA or applicable foreign regulatory agency may suspend or terminate our clinical trials that are initiated, not authorize us to initiate further trials, or if initiated, such further trials could be suspended or terminated. The FDA or comparable foreign regulatory authorities could order us to cease further development of, or deny or withdraw approval of, any of our product candidates for any or all targeted indications.

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

- regulatory authorities may withdraw approvals of such product and require us to take 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 and/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-045, and KPL-404, we were not involved in the development of these product candidates and, as a result, we are dependent on Regeneron, MedImmune, Biogen, Novo Nordisk and Primatope having accurately reported the results and correctly collected and interpreted the data from all pre-clinical and clinical trials conducted prior to our in-license or acquisition.

We had no involvement with or control over the pre-clinical and clinical development of any of our product candidates prior to our in-license or acquisition of them. We are dependent on Regeneron, MedImmune, Biogen, Novo Nordisk, and Primatope having conducted their research and development in accordance with the applicable protocols and legal, regulatory and scientific standards; having accurately reported the results of all pre-clinical studies and clinical trials conducted prior to our in-license or acquisition; and having correctly collected, interpreted, and completely transferred the data from these trials 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 pre-clinical 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 pre-clinical studies, and any positive results we may obtain from our early clinical trials of our product candidates or from the clinical trials conducted by the companies from whom we 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 pre-clinical studies and clinical trials. Similarly, even if we are able to complete our planned pre-clinical studies or clinical trials of our product candidates, the positive results from the pre-clinical studies and clinical trials of our product candidates may not be replicated in our subsequent pre-clinical studies or clinical trial results. 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 late-stage clinical trials after achieving positive results in early-stage development and we cannot be certain that we will not face similar setbacks. These setbacks have been caused by, among other things, pre-clinical findings made while clinical trials were underway or safety or efficacy observations made in pre-clinical studies and clinical trials, including previously unreported adverse events. Moreover, pre-clinical and clinical data are often susceptible to varying interpretations and analyses and many companies that believed their product candidates performed satisfactorily in pre-clinical studies and clinical trials nonetheless failed to obtain FDA or EMA approval. Furthermore, the approval policies or regulations of the FDA or the applicable foreign regulatory agencies may significantly change in a manner rendering our clinical data insufficient for approval, which may lead to the FDA or any foreign regulatory agencies delaying, limiting or denying approval of our product candidates.

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

From time to time, we may publish interim "top-line" or preliminary data from our clinical trials. For example, in December 2018, we released interim data from the open-label Phase 2 proof-of-concept clinical trial of rilonacept in recurrent pericarditis. Preliminary or interim data from clinical trials that we may complete are subject to the risk that one or more of the clinical outcomes may materially change as patient enrollment continues and more patient data become available. Preliminary or interim data also remain subject to audit and verification procedures that may result in the final data being materially different from the preliminary data we previously published. As a result, interim and preliminary data should be viewed with caution until the final data are available. Adverse differences between preliminary or interim data and final data could significantly harm our business prospects.

Regulatory approval processes are lengthy, time consuming and inherently unpredictable. If we are not able to obtain, or if there are delays in obtaining, required regulatory approvals for our product candidates, we will not be able to commercialize, or will be delayed in commercializing, our product candidates and our ability to generate revenue will be materially impaired.

Our product candidates and the activities associated with their development and commercialization, including their design, testing, manufacture, safety, efficacy, recordkeeping, labeling, storage, approval, advertising, promotion, sale, distribution, import and export are subject to comprehensive regulation by the FDA and other regulatory agencies in the United States and by comparable authorities in other countries. Before we can commercialize any of our product candidates, we must obtain marketing approval. We have not received approval or clearance to market any of our product candidates from regulatory authorities in any jurisdiction and it is possible that none of our product candidates or any product candidates we may seek to develop in the future will ever obtain regulatory approval or clearance. We have only limited experience in filing and supporting the applications necessary to gain regulatory approvals and may need to rely on third-party CROs and regulatory consultants to assist us in this process. Securing regulatory approval requires the submission of extensive pre-clinical and clinical data and supporting information to the various regulatory authorities for each therapeutic indication to establish the product candidate's safety and efficacy. Securing regulatory approval also requires the submission of information about the biologic manufacturing process to, and inspection of manufacturing facilities by, the relevant regulatory authority. Our product candidates may not be effective, may be only moderately effective or may prove to have undesirable or unintended side effects, toxicities or other characteristics that may preclude our obtaining marketing approval or prevent or limit commercial use. The FDA and other regulatory authorities have substantial discretion in the approval process, and determining when or whether regulatory approval will be obtained for a product candidate. Even if we believe the data collected from clinical trials are promising, such data may not be sufficient to support approval by the FDA or any other regulatory authority.

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

- the FDA or comparable foreign regulatory authorities may disagree with the number, design or implementation of our clinical trials to support further development or approval;
- we may be unable to demonstrate to the satisfaction of the FDA or comparable foreign regulatory authorities that a product candidate is safe and effective for its proposed indication;
- the results of clinical trials may not meet the level of statistical significance required by the FDA or comparable foreign regulatory authorities for approval;

- we may be unable to demonstrate that a product candidate's clinical and other benefits outweigh its safety risks;
- the FDA or comparable foreign regulatory authorities may disagree with our interpretation of data from pre-clinical studies or clinical trials;
- the 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 evaluate the effectiveness of different doses as well as mavrilimumab's pharmacokinetic profile;
- 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 our product candidates, the commercial prospects for our product candidates may be harmed and our ability to generate revenue will be materially impaired.

Our product candidates regulated as biologics in the United States may face competition sooner than anticipated.

In the United States, the Biologics Price Competition and Innovation Act of 2009, or BPCIA, created an abbreviated approval pathway for biological products that are biosimilar to or interchangeable with an FDA-licensed reference biological product. Under the BPCIA, an application for a biosimilar product may not be submitted to the FDA until four years following the date that the reference product was first licensed by the FDA. In addition, the approval of a biosimilar product may not be made effective by the FDA until 12 years from the date on which the reference product was first licensed. During this 12-year period of exclusivity, another company may still market a competing version of the reference product if the FDA approves a full BLA for the competing product containing the sponsor's own pre-clinical data and data from adequate and well-controlled clinical trials to demonstrate the safety, purity and potency of their product. The law is complex and is still being interpreted and implemented by the FDA. As a result, its ultimate impact, implementation and meaning are subject to uncertainty. While it is uncertain when processes intended to implement the BPCIA may be fully adopted by the FDA, any such processes could have a material adverse effect on the future commercial prospects of our product candidates.

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, 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 European Union, we may not obtain approval or commercialize our product candidates in other markets, which would limit our ability to realize their full market potential.

In order to market any products in a country or territory, we must establish and comply with numerous and varying regulatory requirements of such country or territory regarding safety and efficacy. Clinical trials conducted in one country may not be accepted by regulatory authorities in other countries, and regulatory approval in one country does not mean that regulatory approval will be obtained in any other country. Approval procedures vary among countries and can involve additional product testing and validation and additional administrative review periods. Seeking regulatory approvals in all markets may require additional pre-clinical studies or clinical trials, which would be costly and time consuming. Regulatory requirements can vary widely from country to country and could delay or prevent the introduction of our product candidates in those countries. Satisfying these and other regulatory requirements is costly, time consuming, uncertain and subject to unanticipated delays. In addition, our

failure to obtain regulatory approval in any country may delay or have negative effects on the process for regulatory approval in other countries.

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

As part of our business strategy, we are pursuing 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 European Union, the European Commission grants orphan drug designation after receiving the opinion of the EMA's Committee for Orphan Medicinal Products on an Orphan Drug Designation application. In the European Union, orphan drug designation is intended to promote the development of drugs that are intended for the diagnosis, prevention or treatment of life-threatening or chronically debilitating conditions affecting not more than five in 10,000 persons in the European Union and for which no satisfactory method of diagnosis, prevention, or treatment has been authorized (or the product would be a significant benefit to those affected). Additionally, orphan 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 European Union would be sufficient to justify the necessary investment in developing the drug. In the European Union, orphan drug designation entitles a party to financial incentives such as reduction of fees or fee waivers, as well as potential marketing exclusivity.

In addition, if a drug or biologic with an orphan drug designation subsequently receives the first marketing approval for the indication for which it has such designation, the drug or biologic is entitled to a period of marketing exclusivity, which precludes the FDA from approving another marketing application for the same drug and indication for that time period, except in limited circumstances. If our competitors are able to obtain orphan drug exclusivity prior to 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 European Union. The European Union exclusivity period can be reduced to six years if a drug no longer meets the criteria for orphan drug designation or if the drug is sufficiently profitable so that market exclusivity is no longer justified.

Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent 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 intend to pursue orphan drug designation for certain of our product candidates in addition to rilonacept, 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 pre-clinical data demonstrate the potential to address unmet medical needs for this condition, the sponsor may apply for Fast Track Designation. The FDA has broad discretion whether or not to grant this designation, so even if we believe a particular product candidate is eligible for this designation, we cannot assure you that the FDA would decide to grant it. Even if we have obtained Fast Track Designation for one or more of our product candidates, we may not experience a faster development process, review or approval compared to non-expedited FDA review procedures. In addition, the FDA may withdraw Fast Track Designation 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 meet 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, any of our eventual pivotal trials or related regulatory submissions would prevent us from, or delay us in, obtaining regulatory approval for, or clearance of, our product candidates. In addition, it is possible that the FDA may refuse to accept for substantive review any BLA submissions that we submit for our product candidates or may conclude after review of our applications that they are insufficient to obtain marketing approval or clearance of our product candidates. If the FDA does not accept our applications or issue marketing authorizations for our product candidates, it may require that we conduct additional clinical, pre-clinical or manufacturing validation trials and submit that data before it will reconsider our applications. Depending on the extent of these or any other FDA-required trials, approval of any BLA or receipt of other marketing authorizations for any other applications that we submit may be delayed by several years, or may require us to expend more resources than we have available. It is also possible that additional trials, if performed and completed, may not be considered sufficient by the FDA to approve our BLAs or grant other marketing authorizations. Any delay in obtaining, or an inability to obtain, marketing approvals would prevent us from commercializing our product candidates, generating revenues and achieving and sustaining profitability. If any of these outcomes occur, we may be forced to abandon our development efforts for our product candidates, which could significantly harm our business.

Risks related to manufacturing and our dependence on third parties

We contract with third parties for manufacturing our product candidates and for pre-clinical and clinical development and expect to continue to do so for our commercial supply. This reliance on third parties increases the risk that we may not have sufficient quantities of our product candidates or such quantities at an acceptable cost, which could delay, prevent or impair our development or commercialization efforts.

We do not currently own or operate any manufacturing facilities. Although we are in the process of building small-scale manufacturing facilities to produce drug substance to support certain of our pre-clinical studies and certain of our Phase 1 clinical trials, we rely, and expect to continue to rely, on third parties for the manufacture of our product candidates for the majority of our development efforts, as well as for the commercial manufacture of our product candidates, if approved. We rely on these third parties to develop the processes necessary to produce our product candidates at sufficient quality and quantity to support our development and commercialization efforts. Our reliance increases the risk that we will have insufficient quantities of our product candidates or that our product candidates are not produced at an acceptable cost or quality, which could delay, prevent or impair our development or commercialization efforts.

For example, we contract with Regeneron to produce rilonacept, and with CMOs for the manufacture of KPL-716 drug substance and drug product. Further, we have and plan to continue to enter into agreements with CMOs to produce mavrilimumab beyond our current inventory. We will need to transfer the technology to manufacture mavrilimumab to these CMOs, and these CMOs may be required to adopt different manufacturing protocols or processes. These CMOs will also need to produce mavrilimumab using different

media and feed compared to the processes that were used by MedImmune to develop our existing inventory. We cannot provide any assurance that the technology transfer or process development will be successful, or that any CMO will be successful in producing mavrilimumab in sufficient quantities or of acceptable quality, if at all. In addition, we currently contract with CMOs in connection with certain planned production of our pre-clinical product candidates, KPL-045 and KPL-404, and while we have built small-scale manufacturing facilities to support certain pre-clinical and early clinical development for KPL-045 and KPL-404, 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 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 and/or maintain regulatory approval for their manufacturing facilities. In addition, 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. Further, Regeneron has an exclusive right to produce rilonacept for a period of time.

Any performance failure on the part of our existing or future manufacturers could delay clinical development or marketing approval. If our current CMOs cannot perform as agreed, we may be required to replace such manufacturers. Although we believe that there are several potential alternative manufacturers who could manufacture our product candidates, we may incur added costs and delays in identifying and qualifying any such replacement.

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

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

We built our own small-scale manufacturing facilities to support the early development of our product candidates, KPL-045 and KPL-404, 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 clinical studies for these product candidates.

We built small-scale manufacturing facilities to support pre-clinical and early clinical studies for KPL-045 and KPL-404, as well as other potential product candidates. We may not successfully establish sufficient manufacturing capabilities or 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 and/or interrupt our business operations. Furthermore, environmental laws and regulations are

complex, change frequently and have tended to become more stringent. We cannot predict the impact of such changes and cannot be certain of our future compliance. We do not currently carry biological or hazardous waste insurance coverage.

Manufacturing issues at our planned facilities 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 defects or failures, such as defective products or manufacturing failures. We have limited experience overseeing the manufacturing process of KPL-716 and no experience overseeing the manufacturing process of rilonacept, mavrilimumab, KPL-404 and KPL-045. 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, product defects and other supply disruptions. If microbial, viral or other contaminations are discovered in our product candidates or manufacturing facilities, any related production lot could be lost and the relevant manufacturing facilities may need to close for an extended period of time to investigate and remediate the contaminant. Many additional factors could cause production interruptions at our planned 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 pre-clinical 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. Any delay, interruption or other issues that arise in the manufacture, fill-finish, packaging or storage of our product candidates as a result of a failure of the facilities or operations of third parties to pass any regulatory agency inspection could significantly impair our ability to supply our products and product candidates. Significant noncompliance could also result in the imposition of monetary penalties or other civil or criminal sanctions and damage our reputation.

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

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

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

Moreover, if there is a disruption to one or more of our third-party manufacturers' or suppliers' relevant operations the supply of rilonacept, mavrilimumab or KPL-716 will be delayed until such manufacturer or supplier restores the affected facilities or we or they procure alternative manufacturing facilities or sources of supply. Our ability to progress our pre-clinical and clinical programs could be materially and adversely impacted if any of the third-party suppliers upon which we rely were to experience a significant business challenge, disruption or failure due to issues such as financial difficulties or bankruptcy, issues relating to other customers such as regulatory or quality compliance issues, or other financial, legal, regulatory or reputational issues. Additionally, any damage to or destruction of our third-party manufacturers' or suppliers' facilities or equipment may significantly impair our ability to manufacture our product candidates on a timely basis.

Establishing additional or replacement suppliers for the 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 raw materials required in the manufacture and the formulation of our product candidates are derived from biological sources. Such raw materials are difficult to procure and may be subject to contamination or recall. Access to and supply of sufficient quantities of raw materials which meet the technical specifications for the production process is challenging, and often limited to single-source suppliers. Finding an alternative supplier could take a significant amount of time and involve significant expense due to the nature of the products and the need to obtain regulatory approvals. If we or our manufacturers are unable to purchase the raw materials necessary for the manufacture of our product candidates on acceptable terms, in a timely manner, at sufficient quality levels, or in adequate quantities, if at all, our ability to produce sufficient quantities of our products for clinical or commercial requirements would be negatively impacted. A material shortage, contamination, recall or restriction on the use of certain biologically derived substances or any raw material used in the manufacture of our 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 or cleared.

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

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

We and our CROs will be required to comply with regulations, including GCPs, for conducting, monitoring, recording and reporting the results of clinical trials to ensure that the data and results are scientifically credible and accurate, and that the trial patients are adequately informed of the potential risks of

participating in clinical trials and their rights are protected. These regulations are enforced by the FDA, the Competent Authorities of the Member States of the European Economic Area and comparable foreign regulatory authorities for any products in clinical development. The FDA enforces GCP regulations through periodic inspections of clinical trial sponsors, principal investigators and trial sites. If we or our CROs fail to comply with applicable GCPs, the clinical data generated in our clinical trials may be deemed unreliable and the FDA or comparable foreign regulatory authorities may require us to perform additional clinical trials before approving our marketing applications. We cannot ensure that, upon inspection, the FDA 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 all of the clinical trials. As a result, many important aspects of our development programs, including their conduct and timing, will be outside of our direct control. Our reliance on third parties to conduct future clinical trials will also result in less direct control over the management of data developed through clinical trials than would be the case if we were relying entirely upon our own staff. Communicating with outside parties can also be challenging, potentially leading to mistakes as well as difficulties in coordinating activities. Outside parties may:

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

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

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

If the third parties conducting our pre-clinical studies or our clinical trials do not perform their contractual duties or obligations, experience significant business challenges, disruptions or failures, do not meet

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

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

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

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

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

- collaborators have significant discretion in determining the efforts and resources that they will apply to collaborations;
- a collaborator with marketing, manufacturing and distribution rights to one or more products may not commit sufficient resources to or otherwise not perform satisfactorily in carrying out these activities;
- collaborators may not properly maintain or defend our intellectual property rights or may use our intellectual property or proprietary information in a way that gives rise to actual or threatened litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential liability;

- collaborations may be terminated, and, if terminated, this may result in a need for additional capital to pursue further development or commercialization of the applicable current or future product candidates;
- collaborators may own or co-own intellectual property covering products that results from our collaborating with them, and in such cases, we would not have the exclusive right to develop or commercialize such intellectual property, 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;
- disputes may arise with respect to the ownership of any intellectual property developed pursuant to our collaborations; and
- a collaborator's sales and marketing activities or other operations may not be in compliance with applicable laws resulting in civil or criminal proceedings.

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

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

Risks related to competition, retaining key employees and managing growth

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

The development and commercialization of new drugs and biologics is highly competitive. We face competition with respect to our current product candidates, and will face competition with respect to any product candidates that we may seek to develop or commercialize in the future, from major pharmaceutical companies, specialty pharmaceutical companies and biotechnology companies worldwide. There are a number of large pharmaceutical and biotechnology companies that currently market and sell drugs or biologics 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 currently marketed product that modulates the signaling of IL-1a and

IL-1b, anakinra (KINERET), and one currently marketed product that modulates the signaling of IL-1b, canakinumab (ILARIS). There are other therapies which modulate IL-1a and IL-1b in various stages of clinical development for diseases other than recurrent pericarditis from companies that include AbbVie, Inc., or AbbVie, XBiotech Inc. and Handok Inc. We expect mavrilimumab, if approved, to experience competitive pressure from tocilizumab (ACTEMRA), which was approved in 2017 for use in GCA in combination with glucocorticoids. Additional competition may be experienced from upadacitinib from AbbVie, which is expected to start enrollment for a Phase 3 clinical trial in GCA in December 2018. In addition, Eli Lilly and AbbVie are conducting clinical trials for oral janus kinase inhibitors. Furthermore, Sanofi S.A. and Regeneron intend to initiate a Phase 3 clinical trial with their anti-IL-6 program in 2018, Novartis International AG, plans to start a trial with secukinumab (Cosentyx) and Janssen's ustekinumab (STELARA) is being trialed in two small studies for GCA. KPL-716, if approved for atopic dermatitis, will face competitive pressure from dupilumab (DUPIXENT), which is approved to treat atopic dermatitis. KPL-716 may face additional competition from several systemically administered products currently in development for atopic dermatitis including upadacitinib, PF-04965842, ANB-020, nemolizumab, baricitinib, ASn002, GBR-830, ZPL-389, PF-06817024, MEDI9314, MOR106, ARGX-112, tralokinumab and lebrikizumab. Multiple therapies are in development for prurigo nodularis and any that receive FDA approval for this indication will be likely competitors to KPL-716. These products include nemolizumab, serlopitant and nalbuphine ER.

Many of the companies against which we are competing or against which we may compete in the future have significantly greater financial resources and expertise in research and development, manufacturing, pre-clinical testing, conducting clinical trials, obtaining regulatory approvals and marketing approved products than we do. Mergers and acquisitions in the pharmaceutical, biotechnology and diagnostic industries may result in even more resources being concentrated among a smaller number of our competitors. Smaller or early-stage companies may also prove to be significant competitors, particularly through collaborative arrangements with large and established companies. These competitors also compete with us in recruiting and retaining qualified scientific and management personnel and establishing clinical trial sites and patient registration for clinical trials, as well as in acquiring technologies complementary to, or necessary for, our programs.

Our commercial opportunity could be reduced or eliminated if our competitors develop and commercialize products that are safer, more effective, have fewer or less severe side effects, are more convenient or are less expensive than any drugs that we or our collaborators may develop. Our competitors also may obtain FDA or other regulatory approval for their products more rapidly than we may obtain approval for ours, which could result in our competitors establishing a strong market position before we or our collaborators are able to enter the market. The key competitive factors affecting the success of all of our product candidates, if approved, are likely to be their efficacy, safety, convenience, price, the effectiveness of companion diagnostics in guiding the use of related products, market acceptance by physicians and patients, the level of generic competition and the availability of reimbursement from government and other third-party payors.

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

We have a limited operating history and are highly dependent on the research and development, clinical, commercial and business development expertise of Sanj K. Patel, our Chief Executive Officer and Chairman of the Board of Directors, Stephen Mahoney, our President and Chief Operating Officer, and John F. Paolini, M.D., Ph.D., our Chief Medical Officer, 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, each of them may terminate their employment with us at any time. We do not maintain "key person"

insurance for any of our executives or other employees. In addition, we rely on consultants and advisors, including scientific and clinical advisors, to assist us in formulating our research and development and commercialization strategy. Our consultants and advisors may be employed by employers other than us and may have commitments under consulting or advisory contracts with other entities that may limit their availability to us. If we are unable to continue to attract and retain high quality personnel, our ability to pursue our growth strategy will be limited.

Recruiting and retaining qualified scientific, clinical, manufacturing and sales and marketing personnel 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 and patents relating to KPL-045.

Certain provisions in our intellectual property agreements may be susceptible to multiple interpretations. The resolution of any contract interpretation disagreement that may arise could affect the scope of our rights to the relevant intellectual property or technology, or affect financial or other obligations under the relevant agreement, either of which could have a material adverse effect on our business, financial condition, results of operations and prospects.

We or our licensors have not pursued or maintained, and may not pursue or maintain in the future, patent protection for our products in every country or territory in which we may sell our products. In addition, we cannot be sure that any of our pending patent applications or pending trademark applications will issue or that, if issued, they have or will issue in a form that will be advantageous to us. The United States Patent and Trademark Office, or the USPTO, international patent offices or judicial bodies may deny or significantly narrow claims made under our patent applications and our issued patents may be successfully challenged, may be designed around, or may otherwise be of insufficient scope to provide us with protection for our commercial products. Further, the USPTO, international trademark offices or judicial bodies may deny our trademark applications and, even if published or registered, these trademarks may not effectively protect our brand and goodwill. Like patents, trademarks also may be successfully opposed or challenged.

The patent position of biotechnology and pharmaceutical companies generally is highly uncertain, involves complex legal and factual questions and has in recent years been the subject of much litigation. The degree of patent protection we require to successfully commercialize our product candidates may be unavailable or severely limited in some cases and may not adequately protect our rights or permit us to gain or keep any competitive advantage. We cannot provide any assurances that any of our owned or

in-licensed patents have, or that any of our owned or in-licensed pending patent applications that mature into issued patents will have, claims with a scope sufficient to protect 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 European Union and Japan, subject to the applicable laws in those jurisdictions. We may not receive an extension if we fail to apply within applicable deadlines or fail to apply prior to expiration of relevant patents. For example, no patent term extension was obtained in the United States following the FDA's approval of 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. 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 expect to pursue orphan drug designation for rilonacept in the United States for the treatment of recurrent pericarditis, we may not be successful in obtaining such designation or we may not be able to maintain the benefits of the designation. Even if we obtain orphan drug exclusivity for any of our product candidates, that exclusivity may not effectively protect those product candidates from competition because different drugs can be approved for the same condition, and orphan drug exclusivity does not prevent 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 under which we acquired our product candidates, we could lose the ability to continue the development and commercialization of the related product. Additionally, our current licensing and acquisition agreements contain limitations and restrictions that could limit or adversely affect our ability to develop and commercialize other products in the future.

We entered into agreements to acquire the rights to develop and/or commercialize our product candidates, rilonacept, mavrilimumab, KPL-716, KPL-045 and KPL-404. In September 2017, we entered into a license agreement with Regeneron to obtain an exclusive license under certain intellectual property rights controlled by Regeneron to develop and commercialize 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. Each of these agreements requires us to use commercially reasonable efforts to develop and commercialize the related product candidates, make timely milestone and other payments, provide certain information regarding our activities with respect to such product candidates and indemnify the other party with respect to our development and commercialization activities under the terms of the agreements. In August 2017, we licensed KPL-045 from Novo Nordisk. In September 2017, we entered into a stock and asset purchase agreement with Primatope pursuant to which we have a license to intellectual property controlled by Primatope to research, develop and manufacture KPL-404 and were granted an exclusive license to acquire all of the outstanding capital stock of Primatope. These agreements and any future such agreements that we enter into impose a variety of obligations and related consequences.

We are a party to a number of license and acquisition agreements of importance to our business and to our current product candidates, and we expect to be subject to additional such agreements in the future. Disputes may arise between us and any of these counterparties regarding intellectual property subject to and each parties' obligations under such agreements, including:

- our diligence obligations with respect to the use of the licensed technology in relation to our development and commercialization of our product candidates, and what activities satisfy those diligence obligations;
- the scope of rights granted under the agreement and other interpretation-related issues;
- our obligations to make milestone, royalty or other payments under those agreements;

- whether and the extent to which our technology and processes infringe on intellectual property of the licensor that is not subject to the agreement;
- our right to sublicense patent and other rights to third parties;
- the ownership of inventions and know-how resulting from the joint creation or use of intellectual property by our licensors and us and our partners;
- our right to transfer or assign the license; and
- the effects of termination.

These or other disputes over our obligations or intellectual property that we have licensed or acquired may prevent or impair our ability to maintain our current arrangements on acceptable terms, or may impair the value of the arrangement to us. Any such dispute could have an adverse 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 terminate the relevant agreement. 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.

In January 2019, we exercised our option to acquire Primatope. Subject to a supplemental due diligence period, we expect to close the transaction within 60 days of the option exercise. While we expect to close the transaction, if we fail to do so, our license to the Primatope-controlled KPL-404 intellectual property will terminate, in which case we would cease the development of KPL-404.

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 abandon development of the relevant program or product candidate, which could have a material adverse effect on our business, financial condition, results of operations and prospects.

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

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

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

Competitors and other third parties may infringe, misappropriate or otherwise violate our patents and other intellectual property rights, whether owned or in-licensed. To counter infringement or unauthorized use, we or our current or future collaborators may be required to file infringement claims against these

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

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

An adverse result in any litigation proceeding could put one or more of our patents, whether owned or in-licensed, at risk of being invalidated or interpreted narrowly. If a defendant were to prevail on a legal assertion of invalidity or unenforceability of our patents covering one of our product candidates, we would lose at least part, and perhaps all, of the patent protection covering such product candidate. Competing products may also be sold in other countries in which our patent coverage might not exist or be as strong. If we lose a foreign patent lawsuit, alleging our infringement of a competitor's patents, we could be prevented from marketing our products in one or more foreign countries. Any of these outcomes would have a materially adverse effect on our business.

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

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

We may not have sufficient financial or other resources to adequately conduct such litigation or proceedings. Some of our competitors may be able to sustain the costs of such litigation or proceedings more effectively than we can because of their greater financial resources and more mature and developed intellectual property portfolios. Accordingly, despite our efforts, we may not be able to prevent third

parties from infringing upon or misappropriating or from successfully challenging our intellectual property rights. Uncertainties resulting from the initiation and continuation of patent litigation or other proceedings could have a material adverse effect on our ability to compete in the marketplace.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Risks related to commercialization

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

The precise incidence and prevalence for all the conditions we aim to address with our programs are unknown. Our projections of both the number of people who have these diseases, as well as the subset of people with these diseases who have the potential to benefit from treatment with our product candidates, are based on beliefs and estimates. These estimates have been derived from a variety of sources, including the scientific literature, surveys of clinics, patient foundations or market research, and may prove to be incorrect. Further, new trials may change the estimated incidence or prevalence of these diseases. The number of patients may turn out to be lower than expected.

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

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

We do not currently have a sales, marketing or distribution infrastructure. We have never sold, marketed or distributed any therapeutic products. To achieve commercial success for any approved product candidate, we must build our sales, marketing, managerial and other non-technical capabilities or make arrangements with third parties to perform these services. We currently plan to establish our own sales and marketing capabilities and directly commercialize any approved product candidate.

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

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

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

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

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

Our current or future product candidates may not gain market acceptance by physicians or patients, in which case our ability to generate product revenues will be compromised.

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

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

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

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

Our ability to commercialize any product candidates successfully will depend in part on the extent to which adequate coverage and reimbursement for these product candidates and related treatments will be available from government authorities, private health insurers and other organizations. Government authorities and third-party payors, such as private health insurers and health maintenance organizations, decide which products they will pay for and establish reimbursement levels. A primary trend in the U.S. healthcare industry and elsewhere is cost containment. Government authorities and third-party payors have attempted to control costs by limiting coverage and the amount of reimbursement for particular products. Increasingly, third-party payors are requiring that drug companies provide them with predetermined discounts from list prices and are challenging the prices charged for products. We cannot be sure that adequate coverage will be available for any product candidate that we commercialize and, if coverage is available, that the level of reimbursement will be adequate or that such coverage will not require co-payments that patients may find unacceptably high. Reimbursement may impact the demand for, or the price of, any product candidate for which we obtain marketing approval. If reimbursement is not available or is available only to limited levels, we may not be able to successfully commercialize any product candidate for which we obtain marketing approval. Any coverage or reimbursement that may become available may be decreased or eliminated in the future.

There may be significant delays in obtaining reimbursement for newly approved products, and coverage may be more limited than the purposes for which the product is approved by the FDA or similar regulatory authorities outside the United States. Moreover, eligibility for reimbursement does not imply that any product will be paid for in all cases or at a rate that covers our costs, including research, development, manufacture, sale and distribution. Interim reimbursement levels for new products, if applicable, may also not be sufficient to cover our costs and may not be made permanent. Reimbursement rates may vary according to the use of the product and the clinical setting in which it is used, may be based on reimbursement levels already set for lower-cost products and may be incorporated into existing payments for other services. Net prices for products may be reduced by mandatory discounts or rebates required by government healthcare programs or private payors and by any future relaxation of laws that presently restrict imports of products from countries where they may be sold at lower prices than in the United States. Third-party payors often rely upon Medicare coverage policy and payment limitations in setting their own reimbursement policies. Our inability to promptly obtain coverage and profitable payment rates from both government-funded and private payors for any approved products that we develop could have a material adverse effect on our operating results, our ability to raise capital needed to commercialize drugs and our overall financial condition.

There is significant uncertainty related to the insurance coverage and reimbursement of newly approved products. In the United States, third-party payors, including private and governmental payors, such as the Medicare and Medicaid programs, play an important role in determining the extent to which new drugs and biologics will be covered. The Medicare and Medicaid programs increasingly are used as models in the United States for how private payors and other governmental payors develop their coverage and reimbursement policies for drugs and biologics. Some third-party payors may require pre-approval of coverage for new or innovative drug therapies before they will reimburse healthcare providers who use such therapies. It is difficult to predict at this time what third-party payors will decide with respect to the coverage and reimbursement for our product candidates.

Third-party payors increasingly are challenging prices charged for pharmaceutical or biologic products and services, and many third-party payors may refuse to provide coverage and reimbursement for particular drugs or biologics when an equivalent generic drug, biosimilar or a less expensive therapy is available. It is possible that a third-party payor may consider our product candidates as substitutable and only offer to reimburse patients for the less expensive product. Even if we show improved efficacy or improved convenience of administration with our product candidates, pricing of existing products may limit the amount we will be able to charge for our product candidates. These payors may deny or revoke the reimbursement status of a given product or establish prices for new or existing marketed products at levels that are too low to enable us to realize an appropriate return on our investment in our product candidates. If reimbursement is not available or is available only at limited levels, we may not be able to successfully commercialize our product candidates, and may not be able to obtain a satisfactory financial return on our product candidates.

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

Moreover, increasing efforts by governmental and third-party payors in the United States and abroad to cap or reduce healthcare costs may cause such organizations to limit both coverage and the level of reimbursement for newly approved products and, as a result, they may not cover or provide adequate payment for our product candidates. We expect to experience pricing pressures in connection with the sale of our product candidates due to the trend toward managed healthcare, the increasing influence of health maintenance organizations and additional legislative changes. The downward pressure on healthcare costs in general, particularly prescription drugs and biologics and surgical procedures and other treatments, has become intense. As a result, increasingly high barriers are being erected to the entry of new products.

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

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

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

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

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

If the FDA or a comparable foreign regulatory authority approves any of our product candidates, it will be subject to ongoing regulatory requirements for manufacturing, labeling, packaging, distribution, storage, advertising, promotion, sampling, record-keeping adverse event reporting, conduct of post-marketing trials and submission of safety, efficacy and other post-market information, including both federal and state requirements in the United States and requirements of comparable foreign regulatory authorities.

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

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

We will have to comply with requirements concerning advertising and promotion for our products. Promotional communications with respect to prescription drugs and biologics are subject to a variety of legal and regulatory restrictions and must be consistent with the information in the product's approved label. As such, we may not promote our products for indications or uses for which they do not have approval. The holder of an approved BLA or MAA must submit new or supplemental applications and obtain approval for certain changes to the approved product, product labeling or manufacturing process. We could also be asked to conduct post-marketing clinical trials to verify the safety and efficacy of our products in general or in specific patient subsets. If original marketing approval were obtained via the accelerated approval pathway, we could be required to conduct a successful post-marketing clinical trial to confirm clinical benefit for our products. An unsuccessful post-marketing trial or failure to complete such a trial could result in the withdrawal of marketing approval. The FDA also may place other conditions on approvals including the requirement for a REMS, to assure the safe use of the product. If the FDA concludes a REMS is needed, the sponsor of the BLA must submit a proposed REMS before it can obtain approval. A REMS could include medication guides, physician communication plans or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools.

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

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

Any government investigation of alleged violations of law could require us to expend significant time and resources in response, and could generate negative publicity. Any failure to comply with ongoing regulatory

requirements may significantly and adversely affect our ability to commercialize and generate revenue from our products. If regulatory sanctions are applied or if regulatory approval is withdrawn, the value of our company and our operating results will be adversely affected.

If there are changes in the application of legislation or regulatory policies, or if problems are discovered with a product or the manufacture of a product, or if we or one of our distributors, licensees or co-marketers fails to comply with regulatory requirements, the regulatory authorities could take various actions. These include imposing fines on us, imposing restrictions on our product or its manufacture and requiring us to recall or remove 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, once we begin commercializing our product candidates, if approved, we will be subject to additional healthcare statutory and regulatory requirements and enforcement by the federal government and the states and foreign governments in which we conduct our business. Healthcare providers, physicians and third-party payors will play a primary role in the recommendation and prescription of any product candidates for which we obtain marketing approval. Our future arrangements with third-party payors and customers may expose us to broadly applicable fraud and abuse and other healthcare laws and regulations that may constrain the business or financial arrangements and relationships through which we market, sell and distribute our product

candidates for which we obtain marketing approval. Restrictions under applicable federal and state healthcare laws and regulations, include the following:

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

- the U.S. federal False Claims Act and civil monetary penalties laws, which, among other things, impose criminal and civil penalties, including through civil whistleblower or *qui tam* actions, against individuals or entities for knowingly presenting, or causing to be presented, to the federal government, claims for payment that are false or fraudulent or making a false statement to avoid, decrease or conceal an obligation to pay money to the federal government. In addition, the government may assert that a claim including items and services resulting from a violation of the federal Anti-Kickback Statute constitutes a false 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 health care 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 and/or competent authorities before entering into agreements with physicians.

Ensuring that our future business arrangements with third parties comply with applicable healthcare laws and regulations will involve substantial costs. It is possible that governmental authorities will conclude that our business practices do not comply with current or future statutes, regulations or case law involving applicable fraud and abuse or other healthcare laws and regulations. If our operations, including anticipated activities to be conducted by our sales team, were to be found to be in violation of any of these laws or any other governmental regulations that may apply to us, we may be subject to the imposition of civil, criminal and administrative penalties, damages, disgorgement, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, individual imprisonment, contractual damages, reputational harm, diminished profits and future earnings, additional reporting requirements and/or oversight if we become subject to a corporate integrity agreement or similar agreement to resolve allegations of non-compliance with these laws, and curtailment or restructuring of our operations, any of which could adversely affect our ability to operate our business and our results of operations. Further, defending against any such actions can be costly, time consuming and may require significant personnel resources. Therefore, even if we are successful in defending against any such actions that may be brought against us, our business may be impaired.

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

We will face an inherent risk of product liability exposure related to the testing of our product candidates in human clinical trials and will face an even greater risk if we commercially sell any product candidates that we may develop. If we cannot successfully defend ourselves against claims that our product candidates caused injuries, we could incur substantial liabilities. Regardless of merit or eventual outcome, liability claims may result in:

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

Although we maintain product liability insurance coverage, it may not be adequate to cover all liabilities that we may incur and is subject to deductibles and coverage limitations. We anticipate that we will need to increase our insurance coverage when and if we successfully commercialize any product candidate. Insurance coverage is increasingly expensive. We may not be able to maintain insurance coverage at a reasonable cost or in an amount adequate to satisfy any liability that may arise. If we are unable to obtain insurance at acceptable cost or otherwise protect against potential product liability claims, we will be exposed to significant liabilities, which may materially and adversely affect our business and financial position. These liabilities could prevent or interfere with our commercialization efforts.

Other risks related to our business

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 and 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 assure you 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 technology and/or businesses often require significant payments and

expenses and will consume additional resources. We will need to continue to devote a substantial amount of time and personnel to research, develop and commercialize any in-licensed or acquired technology, or integrate any new business, in addition to our efforts on our existing portfolio of programs. 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 and/or acquire businesses or undertake business combinations, collaborations, or other strategic transactions;
- for product candidates we seek to in-license or acquire and/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;
- our product candidates may not succeed in pre-clinical studies or clinical trials;
- we may not succeed in formulation or process development;
- any product candidates to which we acquire the rights or that we discover 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 parties' 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, and/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, and/or cause dilution to our existing shareholders if we issue equity securities for all or a portion of the consideration. For example, we exercised our option to acquire all of the outstanding capital stock of Primatope in January 2019, and, subject to a supplemental due diligence period, we expect to close the transaction within 60 days of the option exercise. If we close the transaction, we will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10.0 million as well as potential milestone payments of up to $8.0 million ($3.0 million of which has been achieved as of the date of this prospectus and would be payable at closing), payable in a combination of cash and our Class A common shares. If we do not close the transaction, our license to intellectual property controlled by Primatope to research, develop and manufacture KPL-404 will terminate, in which case we would cease the development of KPL-404. 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 for the transaction.

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

In the United States, European Union and other jurisdictions, there have been and we expect there will continue to be a number of legislative and regulatory initiatives and proposed changes to the healthcare system that could affect our future operations. For example, in the United States, 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, including our product candidates, and a new Medicare Part D coverage gap discount program, in which manufacturers must agree to offer 50% point-of-sale discounts which, through subsequent legislative amendments, was increased to 70%, off negotiated prices of applicable brand drugs and biologics to eligible beneficiaries during their coverage gap period, as a condition for the manufacturer's outpatient products to be covered under Medicare Part D.

Since its enactment, there have been judicial and Congressional challenges to certain aspects of the Affordable Care Act. The current Presidential Administration and U.S. Congress have attempted and will likely continue to seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Most recently, the Tax Cuts and Jobs Act was enacted, which, among other things, removes penalties for not complying with the Affordable Care Act's individual mandate to carry health insurance. 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 U.S. 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 or put pressure on our product pricing.

In the European Union, similar political, economic and regulatory developments may affect our ability to profitably commercialize our product candidates, if approved. In addition to continuing pressure on prices and cost containment measures, legislative developments at the EU or member state level may result in significant additional requirements or obstacles that may increase our operating costs. The delivery of healthcare in the European Union, including the establishment and operation of health services and the pricing and reimbursement of medicines, is almost exclusively a matter for national, rather than EU, law and policy. National governments and health service providers have different priorities and approaches to the delivery of 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 European Union, reimbursement and healthcare payment systems vary significantly by country, and many countries have instituted price ceilings on specific products and therapies.

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

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

Our results of operations could be adversely affected by general conditions in the global economy and in the global financial markets. For example, the global financial crisis caused extreme volatility and disruptions in the capital and credit markets. A severe or prolonged economic downturn, such as the global financial crisis, could result in a variety of risks to our business, including, weakened demand for our product candidates and our ability to raise additional capital when needed on acceptable terms, if at all. A weak or declining economy could also strain our suppliers, possibly resulting in supply disruption, or cause our customers to delay making payments for our services. Doing business internationally involves a number of risks, including but not limited to:

- multiple, conflicting and changing laws and regulations such as privacy regulations, tax laws, export and import restrictions;
- employment laws, regulatory requirements and other governmental approvals, permits and licenses;
- failure by us to obtain and maintain regulatory approvals for the use of our products in various countries;
- additional potentially relevant third-party patent rights;
- complexities and difficulties in obtaining protection and enforcing our intellectual property;
- difficulties in staffing and managing foreign operations;
- complexities associated with managing multiple payor reimbursement regimes, government payors or patient self-pay systems;
- limits in our ability to penetrate international markets;
- financial risks, such as longer payment cycles, difficulty collecting accounts receivable, the impact of local and regional financial crises on demand and payment for our products and exposure to foreign currency exchange rate fluctuations;
- natural disasters, political and economic instability, including wars, terrorism, political unrest, outbreak of disease and boycotts;
- curtailment of trade, and other business restrictions;
- certain expenses including, among others, expenses for travel, translation and insurance; and

- regulatory and compliance risks that relate to maintaining accurate information and control over sales and activities that may fall within the purview of the U.S. Foreign Corrupt Practices Act, its books and records provisions or its anti-bribery provisions.

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

Our internal computer systems, or those of our third-party CMOs, CROs or other contractors or consultants, may fail or suffer security breaches, which could result in a material disruption of our product candidates' development programs.

Despite the implementation of security measures, our internal computer systems and those of our third-party CMOs, CROs and other contractors and consultants are vulnerable to damage from computer viruses, unauthorized access, theft, natural disasters, terrorism, war and telecommunication and electrical failures. While we have not experienced any such system failure, accident or security breach to date, if such an event were to occur and cause interruptions in our operations, it could result in a material disruption of our programs. For example, the loss of clinical trial data for our product candidates could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any disruption or security breach results in a loss of or damage to our data or applications or other data or applications relating to our technology or product candidates, or inappropriate disclosure or theft of confidential or proprietary information, we could incur liabilities and the further development of our product candidates could be delayed.

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

Most healthcare providers, including research institutions from which we obtain patient health information, are subject to privacy and security regulations promulgated under HIPAA, as amended by the HITECH. 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 European Union into the United States may result in the imposition of criminal and administrative sanctions on such collaborators, which could adversely affect our business.

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

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

The EU's new 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 European Union and became binding across all EU member states. The GDPR is in the process of taking effect in the European Economic Area, or 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 Privacy Shield Framework and the standard contractual clauses), and our work 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 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 may knowingly or inadvertently make use of social media in ways that may not comply with our social media policy or other legal or contractual requirements, which may give rise to liability, lead to the loss of trade secrets or other intellectual property or result in public exposure of personal information of our employees, clinical trial patients, customers and others. Furthermore, negative posts or comments about us or our product candidates in social media could seriously damage our reputation, brand image and goodwill. Any of these events could have a material adverse effect on our business, prospects, operating results and financial condition and could adversely affect the price of our Class A common shares.

Our employees, principal investigators, CROs, consultants and other third-party service providers may engage in misconduct or other improper activities, including 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 and/or negligent conduct or disclosure of unauthorized activities to us that violate the regulations of the FDA and other regulatory authorities, including those laws requiring the reporting of true, complete and accurate information to such authorities; healthcare fraud and abuse laws and regulations in the United States and abroad; or laws that require the reporting of financial information or data accurately.

In particular, sales, marketing and business arrangements in the healthcare industry are subject to extensive laws and regulations intended to prevent fraud, misconduct, kickbacks, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, including off-label promotion, sales commission, customer incentive programs and other business arrangements. Activities subject to these laws also involve the improper use of information obtained in the course of clinical trials or creating fraudulent data in our pre-clinical studies or clinical trials, which could result in regulatory sanctions and cause serious harm to our reputation.

We 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 and this offering

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 all matters submitted to our shareholders for approval. This concentration of ownership of our Class B common shares 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.

Our Class A1 common shares and Class B1 common shares have no voting rights. As a result, all matters submitted to our shareholders will be decided by the vote of holders of our Class A common shares and Class B common shares. Each Class A common share is entitled to one vote per Class A common share and each Class B common share is entitled to ten votes per Class B common share. 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 all matters submitted to our shareholders for approval. Immediately following this offering and the concurrent private placement, the holders of Class A common shares will account for 28.5% of our aggregate voting power and the holders of Class B common shares will account for the remaining 71.5% of our aggregate voting power. As a result of the Class A common shares and Class B common shares they hold, our executive officers and certain other members of our senior management will hold 66.8% of our voting power immediately following this offering and the concurrent private placement and will continue to have the ability to control the outcome of all matters submitted to our shareholders for approval. This concentrated control limits other shareholders' ability to influence corporate matters and may have an adverse effect on the price of our Class A common shares, including our Class A common shares being undervalued. Holders of our Class B common shares collectively control our management and affairs and the outcome of matters submitted to our shareholders for approval, including the election of directors. These holders may have interests, with respect to their investment, that are different from our other shareholders, including the investors in this offering. In addition, this concentration of ownership 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.

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 B1 common shares has the ability to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time, and each holder of our Class A1 common shares has the ability to convert any portion of its Class A1 common shares into Class A common shares at any time. 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 holders provide us with 61-days' prior notice that they intend to increase, decrease or waive such threshold upon conversion. Due to these conversion rights, holders of our Class A1 common shares and our Class B1 common shares could, at any time, significantly increase their voting control of us, which could result in their ability to significantly

influence or control matters submitted to our shareholders for approval. 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 those individual holders of Class B common shares who retain their shares in the long term. In addition, such conversion would decrease the ability of the current holders of our Class B common shares, Class A1 common shares or our Class B1 common shares to significantly influence or control matters submitted to our shareholders for approval.

Following the concurrent private placement, entities managed by Baker Brothers will hold 65.0% of our Class A1 common shares and 100% of our Class B1 common shares. Upon 61-days' prior written notice, these entities could convert their Class A1 common shares and Class B1 common shares into Class A common shares and Class B common shares, which in the aggregate would result in such entities holding over 70% of the voting power of our outstanding share capital following this offering and the concurrent private placement. See the sections titled "Description of share capital—Common shares" and "Description of share capital—Voting rights" for more information about the conversion and voting rights associated with our common shares.

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

Our share price is likely to be volatile. The 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, you may not be able to sell your Class A common shares at or above the offering price. The market price for our Class A common shares may be influenced by many factors, including:

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

- changes in the structure of healthcare payment systems;
- market conditions in the pharmaceutical and biotechnology sectors;
- general economic, industry and market conditions;
- changes in voting control of our 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.

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 substantial number of our Class A common shares in the public market could cause our share price to fall.

If our existing shareholders sell, or indicate an intention to sell, substantial amounts of our Class A common shares in the public market after the lock-up and other legal restrictions on resale discussed in this prospectus lapse, the market price of our Class A common shares could decline. Based upon the number of common shares outstanding as of December 31, 2018, upon the completion of this offering and the concurrent private placement, we will have outstanding a total of 18,452,204 Class A common shares, 4,638,855 Class B common shares, 14,995,954 Class A1 common shares and 16,057,618 Class B1 common shares, assuming no exercise of options to purchase Class A common shares outstanding as of December 31, 2018 and no exercise of the underwriters' option to purchase additional Class A common shares. All of the Class A common shares, including Class A common shares issuable upon conversion of our Class B common shares, Class A1 common shares and Class B1 common shares, will be freely tradable without restriction in the public market immediately following this offering and the concurrent private placement, other than shares that are subject to lock-up agreements in connection with this offering and shares that are subject to restrictions on sales under the rules of the Securities Act.

Our directors and executive officers have entered into lock-up agreements with the underwriters of this offering, which restrict their ability to sell or transfer their common shares for 90 days following the date of this prospectus, subject to certain exceptions. After the lock-up agreements expire, up to an additional 6,737,560 Class A common shares held by our directors and executive officers will be eligible for sale in the public market (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares). However, these shares will remain subject to certain limitations on sales made by affiliates pursuant to Rule 144 under the Securities Act for so long as the holders are affiliates for purposes of Rule 144.

In addition, our Class B common shares and Class B1 common shares automatically convert into Class A common shares upon transfer to non-affiliates. As a result, as of December 31, 2018, up to 20,696,473 of our Class A common shares may be issued upon such transfers and may become eligible for sale in the

public market, subject to any lock-up agreements and Rule 144 under the Securities Act. As of December 31, 2018, there were also 5,960,939 of our 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 various vesting schedules, any lock-up agreements and Rule 144 and Rule 701 under the Securities Act. If any of these additional Class A common shares are sold, or if it is perceived that they will be sold, in the public market, the market price of our Class A common shares could decline.

Following this offering and the concurrent private placement, certain holders of our common shares are entitled to rights with respect to the registration of up to 37,670,093 Class A common shares (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares), subject, as applicable, to the lock-up agreements described above. Registration of these shares under the Securities Act would result in the shares becoming freely tradable without restriction under the Securities Act, except for shares purchased by affiliates. Any sales of securities by these shareholders could have a material adverse effect on the market price of our Class A common shares.

If you purchase Class A common shares in this offering, you will suffer immediate dilution of your investment.

The offering price of our Class A common shares is substantially higher than the as adjusted net tangible book value per common share. Therefore, if you purchase Class A common shares in this offering, you will pay a price per Class A common share that substantially exceeds our as adjusted net tangible book value per common share after this offering and the concurrent private placement. To the extent outstanding options are exercised, you will incur further dilution. Based on the offering price of $18.26 per share, you will experience immediate dilution of $10.90 per common share, representing the difference between our as adjusted net tangible book value per common share after giving effect to this offering and the concurrent private placement and the offering price. See "Dilution."

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

We will need additional capital in the future to continue our planned operations. To the extent we raise additional capital by issuing 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. If we sell common shares, convertible securities or other equity securities in more than one transaction, investors may be materially diluted by subsequent sales. These sales may also result in material dilution to our existing shareholders, and new investors could gain rights superior to our existing shareholders.

We have broad discretion in how we use the proceeds of this offering and the concurrent private placement and may not use these proceeds effectively, which could affect our results of operations and cause our share price to decline.

We will have considerable discretion in the application of the net proceeds of this offering and the concurrent private placement. We intend to use the net proceeds from this offering and the concurrent private placement, together with our existing cash and cash equivalents, to advance the clinical development of rilonacept, mavrilimumab and KPL-716, and for working capital and other general

corporate purposes. We may also use a portion of the net proceeds from this offering and the concurrent private placement to in-license, acquire or invest in additional businesses, technologies, products or assets. As a result, investors will be relying upon management's judgment with only limited information about our specific intentions for the use of the balance of the net proceeds of this offering and the concurrent private placement. We may use the net proceeds for purposes that do not yield a significant return or any return at all for our shareholders. In addition, pending their use, we may invest the net proceeds from this offering and the concurrent private placement in a manner that does not produce income or that loses value.

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 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 (a) the last day of the fiscal year in which we have total annual gross revenues of $1.07 billion or more; (b) the last day of the fiscal year following the fifth anniversary of the date of the completion of our IPO; (c) the date on which we have issued more than $1 billion in nonconvertible debt during the previous three years; or (d) the date on which we are deemed to be a large accelerated filer under the rules of the Securities and Exchange Commission, which means the market value of our 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 this prospectus, we have not included all of the executive compensation related information that would be required if we were not an emerging growth company.

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 the fiscal year following the determination that our voting and non-voting common shares held by non-affiliates is more than $250 million measured on the last business day of our second fiscal quarter, or our annual revenues are more than $100 million during the most recently completed fiscal year and our voting and non-voting common shares held by non-affiliates is more 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, are exempt from the auditor attestation requirements of Section 404, 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 our shares price 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 that we did not incur as a private company. In addition, the Sarbanes-Oxley Act of 2002 and rules subsequently implemented by the Securities and Exchange Commission and 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 will increase our legal and financial compliance costs and will make some activities more time-consuming and costly, particularly after we are no longer an emerging growth company and a smaller reporting company. For example, we expect that these rules and regulations may make it more difficult and more expensive for us to obtain director and officer liability insurance.

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

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

Our amended and restated bye-laws contain provisions that could make it more difficult for a third party to acquire us without the consent of our board of directors. These provisions 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, Class B common shares, Class A1 common shares, and Class B1 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. See "Description of share capital."

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

We have never declared or paid cash dividends on our 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 your sole source of gain for the foreseeable future.

Risks related to owning shares in a Bermuda exempted company and certain tax risks

We are a Bermuda company and it may be difficult for 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. See "Enforcement of civil liabilities under United States federal securities laws" for additional information.

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 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 Bermuda Companies Act 1981, as amended, or the Companies Act, or out of or in connection with our bye-laws, including any question regarding the existence and scope of any bye-law and/or whether there has been a breach of the Companies Act or the 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 circumstance 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 a subsidiary in the United States. If we succeed in growing our business, we expect to conduct increased operations through our subsidiaries in various tax jurisdictions pursuant to transfer pricing arrangements between us and our subsidiaries. If two or more affiliated companies are located in different countries, the tax laws or regulations of each country generally will require that transfer prices be the same as those between unrelated companies dealing at arms' length and that appropriate documentation is maintained to support the transfer prices. While we believe that we operate in compliance with applicable transfer pricing laws and intend to continue to do so, our transfer pricing procedures are not binding on applicable tax authorities.

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

Changes in our effective tax rate may reduce our net income in future periods.

Our tax position could be adversely impacted by changes in tax rates, tax laws, tax practice, tax treaties or tax regulations or changes in the interpretation thereof by the tax authorities in Europe (including the United Kingdom), the United States, Bermuda and other jurisdictions, as well as being affected by certain changes currently proposed by the Organization for Economic Co-operation and Development and their action plan on Base Erosion and Profit Shifting. Such changes may become more likely as a result of recent economic trends in the jurisdictions in which we operate, particularly if such trends continue. If such a situation was to arise, it could adversely impact our tax position and our effective tax rate. Failure to manage the risks associated with such changes, or misinterpretation of the laws providing such changes, could result in costly audits, interest, penalties and reputational damage, which could adversely affect our business, results of our operations and our financial condition.

Our actual effective tax rate may vary from our expectation and that variance may be material. A number of factors may increase our future effective tax rates, including:

- the jurisdictions in which profits are determined to be earned and taxed;
- the resolution of issues arising from any future tax audits with various tax authorities;
- changes in the valuation of our deferred tax assets and liabilities;
- increases in expenses not deductible for tax purposes, including transaction costs and impairments of goodwill in connection with acquisitions;
- changes in the taxation of share-based compensation;
- changes in tax laws or the interpretation of such tax laws, and changes in generally accepted accounting principles; and
- challenges to the transfer pricing policies related to our structure.

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

Because we do not expect to earn revenue from our business operations during the current taxable year, and because our sole source of income currently is interest on bank accounts held by us, we believe we will likely be classified as a "passive foreign investment company," or PFIC, for the current taxable year. A non-U.S. company will be considered a PFIC for any taxable year if (i) at least 75% of its gross income is passive income (including interest income), or (ii) at least 50% of the value of its assets (based on an average of the quarterly values of the assets during a taxable year) is attributable to assets that produce or are held for the production of passive income. If we are classified as a PFIC in any year with respect to which a U.S. Holder 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 (1) is subject to the supervision of a U.S. court and the control of one or more "United States persons" (within the meaning of Section 7701(a)(30) of the U.S. Internal Revenue Code of 1986, as amended), or (2) 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. See "Material Bermuda and U.S. federal income tax considerations —Material U.S. federal income tax considerations to U.S. holders—Passive foreign investment company."

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

We believe we are classified as a controlled foreign corporation for the current taxable year and may be classified as a controlled foreign corporation in future taxable years. Even if we were not classified as a controlled foreign corporation, if our group includes one or more U.S. subsidiaries, certain of our non-U.S. subsidiaries could be treated as controlled foreign corporations. If a U.S. Holder 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 we or any of our non-U.S. subsidiaries, if any, are treated as a controlled foreign corporation or whether such investor is treated as a United States shareholder with respect to any of such controlled foreign corporations. Further, we cannot provide any assurances that we will furnish to any United States shareholders information that may be necessary to comply with the reporting and tax paying obligations discussed above. U.S. Holders should consult their tax advisors regarding the potential application of these rules to any investment in our common shares.

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

The U.S. government has recently enacted comprehensive tax legislation that includes significant changes to the taxation of business entities, referenced herein as the Tax Reform Act. These changes include, among others, a permanent reduction to the corporate income tax rate, 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.

Special note regarding forward-looking statements

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

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

In some cases, you can identify forward-looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "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 prospectus are only predictions. We have based these forward-looking statements largely on our current expectations and projections about future events and financial trends that we believe may affect our business, financial condition and results of operations. These forward-looking statements speak only as of the date of this prospectus and are subject to a number of risks, uncertainties and assumptions described under the sections in this prospectus entitled "Risk factors" and "Management's discussion and analysis of financial condition and results of operations" and elsewhere in this prospectus. These forward-looking statements are subject to numerous risks, including, without limitation, the following:

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

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

Because forward-looking statements are inherently subject to risks and uncertainties, some of which cannot be predicted or quantified and some of which are beyond our control, you should not rely on these forward-looking statements as predictions of future events. The events and circumstances reflected in our forward-looking statements may not be achieved or occur and actual results could differ materially from those projected in the forward-looking statements. Moreover, we operate in an evolving environment. New risk factors and uncertainties may emerge from time to time, and it is not possible for management to predict all risk factors and uncertainties. As a result of these factors, we cannot assure you that the forward-looking statements in this prospectus will prove to be accurate. Except as required by applicable law, we do not plan to publicly update or revise any forward-looking statements contained herein, whether as a result of any new information, future events, changed circumstances or otherwise.

You should read this prospectus and the documents that we reference in this prospectus and have filed as exhibits to the registration statement, of which this prospectus is a part, completely and with the understanding that our actual future results may be materially different from what we expect. We qualify all of our forward-looking statements by these cautionary statements.

Industry and other data

Unless otherwise indicated, certain industry data and market data included in this prospectus were obtained from independent third-party surveys, market research, publicly available information, reports of governmental agencies and industry publications and surveys. All of the market data used in this prospectus involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. We believe that the information from these industry publications and surveys included in this prospectus is reliable. The industry in which we operate is subject to a high degree of uncertainty and risk due to a variety of factors, including those described in the sections of this prospectus entitled "Risk factors" and "Special note regarding forward-looking statements" and elsewhere in this prospectus. These and other factors could cause results to differ materially from those expressed in the estimates made by the independent parties and by us.

Use of proceeds

We estimate that the net proceeds to us from our issuance and sale of 2,654,984 Class A common shares in this offering will be approximately $44.8 million (or $51.7 million if the underwriters exercise in full their option to purchase additional Class A common shares) and that the net proceeds to us from our issuance and sale of 2,000,000 Class A1 common shares in the concurrent private placement will be approximately $34.5 million, each based on the offering price of $18.26 per share, and after deducting the underwriting discounts and commissions, placement agent fees and estimated offering expenses payable by us.

We intend to use the net proceeds from this offering and the concurrent private placement, together with our existing cash resources, to advance the clinical development of, and commercial preparation for, rilonacept, to advance the clinical development of mavrilimumab and KPL-716, and the remainder to fund new and ongoing research and development activities, including to advance the pre-clinical development of KPL-045 and KPL-404, and for working capital and other general corporate activities.

This expected use of net proceeds from this offering and the concurrent private placement represents our current intentions based upon our current plans and business conditions. As of the date of this prospectus, we cannot predict with complete certainty all of the particular uses for the net proceeds from this offering and the concurrent private placement or the actual amounts that we will spend on the uses set forth above. We may also use a portion of the net proceeds to in-license, acquire or invest in additional businesses, technologies, products or assets, although currently we have no specific agreements, commitments or understandings in this regard. The amounts and timing of our actual expenditures will depend on numerous factors, including the progress of our clinical trials, our ability to obtain marketing approval from the FDA for our product candidates and other development and commercialization efforts for our product candidates, as well as the amount of cash used in our operations. We may find it necessary or advisable to use the net proceeds from this offering and the concurrent private placement for other purposes, and as a result, our management will retain broad discretion over the allocation of the net proceeds from this offering and the concurrent private placement.

We anticipate that our existing cash, cash equivalents and short-term investments, together with the anticipated net proceeds from this offering and the concurrent private placement, will be sufficient to fund our operating expenses and capital expenditure requirements into 2021. We have based this estimate on assumptions that may prove to be wrong, and we could utilize our available capital resources sooner than we expect. Following this offering and the concurrent private placement, we will require substantial capital to complete clinical development, seek regulatory approval of, and, if approved, commercialize our product candidates.

Pending the use of the proceeds described above, we plan to invest the net proceeds from this offering and the concurrent private placement in short- and intermediate-term, interest-bearing obligations, investment-grade instruments, certificates of deposit or direct or guaranteed obligations of the U.S. government.

Dividend policy

We have never declared or paid any cash dividends on our common shares. In October 2015, we distributed Class B common shares to the then-existing holders of our Class A common shares on a pro rata basis. We intend to retain all of our future earnings, if any, to finance the operation and expansion of our business and do not anticipate paying any cash dividends in the foreseeable future. Any future determination to declare and pay dividends to holders of our common shares will be made at the discretion of our board of directors, which may take into account several factors, including general economic conditions, our financial condition and results of operations, available cash and current and anticipated cash needs, capital requirements, contractual, legal, tax and regulatory restrictions, the implications of the payment of dividends by us to our shareholders and any other factors that our board of directors may deem relevant. In addition, pursuant to the Companies Act, a company may not declare or pay dividends if there are reasonable grounds for believing that (1) the company is, or would after the payment be, unable to pay its liabilities as they become due or (2) that the realizable value of its assets would thereby be less than its liabilities. Under our amended and restated bye-laws, each of our common shares is entitled to dividends if, as and when dividends are declared by our board of directors, subject to any preferred dividend right of the holders of any preferred shares.

Capitalization

The following table sets forth our cash, cash equivalents and short-term investments and our capitalization as of September 30, 2018:

- on an actual basis; and
- on an as adjusted basis to give further effect to our issuance and sale of 2,654,984 Class A common shares in this offering and our issuance and sale of 2,000,000 Class A1 common shares in the concurrent private placement, each at the offering price of $18.26 per share, after deducting the underwriting discounts and commissions, placement agent fees and estimated offering expenses payable by us.

You should read the following table in conjunction with our consolidated financial statements and the related notes appearing at the end of this prospectus and the sections of this prospectus titled "Selected consolidated financial data," "Management's discussion and analysis of financial condition and results of operations" and "Description of share capital."

	As of September 30, 2018	
	Actual	As Adjusted
	(in thousands, except share and per share data)	
Cash, cash equivalents and short-term investments	$ 337,863	$ 417,188
Shareholders' equity:		
Class A common shares, $0.000273235 par value; 15,772,257 shares issued and outstanding, actual; 18,427,241 shares issued and outstanding, as adjusted	$ 4	$ 5
Class B common shares, $0.000273235 par value; 4,638,855 shares issued and outstanding, actual; 4,638,855 shares issued and outstanding, as adjusted	1	1
Class A1 common shares, $0.000273235 par value; 12,995,954 shares issued and outstanding, actual; 14,995,954 shares issued and outstanding, as adjusted	4	4
Class B1 common shares, $0.000273235 par value; 16,057,618 shares issued and outstanding, actual; 16,057,618 shares issued and outstanding, as adjusted	4	4
Additional paid-in capital	470,600	549,924
Accumulated other comprehensive income	(55)	(55)
Accumulated deficit	(151,645)	(151,645)
Total shareholders' equity	318,913	398,238
Total capitalization	$ 318,913	$ 398,238

The foregoing table excludes:

- 5,860,168 Class A common shares issuable upon exercise of share options outstanding as of September 30, 2018, at a weighted average exercise price of $9.78 per share;

- 3,280,059 Class A common shares available for future issuance under our 2018 Plan as of September 30, 2018, as well as common shares that become available pursuant to provisions in the 2018 Plan that automatically increase the share reserve under the 2018 Plan as described in "Executive and director compensation—Executive compensation plans—2018 incentive award plan"; and

- 670,000 Class A common shares available for future issuance under our ESPP as of September 30, 2018, as well as common shares that become available pursuant to provisions in the ESPP that automatically increase the share reserve under the ESPP as described in "Executive and Director Compensation—Executive Compensation Plans—2018 Employee Share Purchase Plan".

Dilution

If you invest in our Class A common shares in this offering, your ownership interest will be immediately diluted to the extent of the difference between the offering price per share and the as adjusted net tangible book value per common share after this offering.

Our historical net tangible book value as of September 30, 2018 was $318.9 million, or $6.45 per common share. Our historical net tangible book value represents our total tangible assets less our total liabilities. Historical net tangible book value per share represents historical net tangible book value divided by the 49,464,684 common shares outstanding as of September 30, 2018.

After giving effect to our issuance and sale of 2,654,984 Class A common shares in this offering and our issuance and sale of 2,000,000 Class A1 common shares in the concurrent private placement, each at the offering price of $18.26 per share, and after deducting the underwriting discounts and commissions, placement agent fees and estimated offering expenses payable by us, our as adjusted net tangible book value as of September 30, 2018 would have been $398.2 million, or $7.36 per share. This represents an immediate increase in as adjusted net tangible book value of $0.91 per share to our existing shareholders and immediate dilution of $10.90 per share to new investors purchasing Class A common shares in this offering and Class A1 common shares in the concurrent private placement. Dilution per share to new investors is determined by subtracting the as adjusted net tangible book value per share after this offering and the concurrent private placement from the offering price per common share paid by new investors. The following table illustrates this dilution on a per share basis:

Public offering price per share		$ 18.26
Historical net tangible book value per share as of September 30, 2018	$ 6.45	
Increase in as adjusted net tangible book value per share attributable to new investors purchasing Class A common shares in this offering and Class A1 common shares in the concurrent private placement	0.91	
As adjusted net tangible book value per share after this offering and the concurrent private placement		7.36
Dilution per share to new investors purchasing Class A common shares in this offering and Class A1 common shares in the concurrent private placement		$ 10.90

If the underwriters exercise in full their option to purchase additional Class A common shares, the as adjusted net tangible book value per share after this offering and the concurrent private placement would be $7.43, and the dilution per share to new investors would be $10.83, in each case based on the offering price of $18.26 per share, and after deducting the underwriting discounts and commissions.

The following table summarizes, on the as adjusted basis as described above, the total number of common shares purchased from us, the total consideration paid and the average price per share paid or to be paid by existing shareholders and by new investors acquiring our Class A common shares in this offering and our Class A1 common shares in the concurrent private placement, each at the offering price of $18.26 per

share, before deducting the underwriting discounts and commissions, placement agent fees and estimated offering expenses payable by us.

	Shares purchased		Total consideration		Average price per share
	Number	Percent	Amount	Percent	
Existing shareholders	49,464,684	91.4%	$ 320,028,545	79.0%	$ 6.47
New investors	4,654,984	8.6	85,000,008	21.0	$ 18.26
Total	54,119,668	100.0%	$ 405,028,553	100.0%	

The table above assumes no exercise of the underwriters' option to purchase additional Class A common shares. If the underwriters exercise in full their option to purchase additional Class A common shares, the percentage of our common shares held by existing shareholders would be decreased to 90.7% of the total number of our common shares outstanding after this offering and the concurrent private placement, and the number of shares held by new investors participating in this offering and the concurrent private placement would be increased to 9.3% of the total number of our common shares outstanding after this offering and the concurrent private placement.

The foregoing tables exclude:

- 5,860,168 Class A common shares issuable upon exercise of share options outstanding as of September 30, 2018, at a weighted average exercise price of $9.78 per share;
- 3,280,059 Class A common shares available for future issuance under our 2018 Plan as of September 30, 2018, as well as common shares that become available pursuant to provisions in the 2018 Plan that automatically increase the share reserve under the 2018 Plan as described in "Executive and director compensation—Executive compensation plans—2018 incentive award plan"; and
- 670,000 Class A common shares available for future issuance under our ESPP as of September 30, 2018, as well as common shares that become available pursuant to provisions in the ESPP that automatically increase the share reserve under the ESPP as described in "Executive and director compensation—Executive compensation plans—2018 employee share purchase plan".

To the extent any of the outstanding share options are exercised, you will experience further dilution as a new investor in this offering. In addition, we may choose to raise additional capital because of market conditions or strategic considerations, even if we believe that we have sufficient funds for our current or future operating plans. If we raise additional capital through the sale of equity or convertible debt securities, the issuance of these securities could result in further dilution to our shareholders.

Selected consolidated financial data

You should read the following selected consolidated financial data together with our consolidated financial statements and the related notes appearing at the end of this prospectus and the "Management's discussion and analysis of financial condition and results of operations" section of this prospectus. We have derived the consolidated statement of operations data for the years ended December 31, 2016 and 2017 and the consolidated balance sheet data as of December 31, 2016 and 2017 from our audited consolidated financial statements appearing at the end of this prospectus. We have derived the consolidated statement of operations data for the nine months ended September 30, 2017 and 2018 and the consolidated balance sheet data as of September 30, 2018 from our unaudited consolidated financial statements appearing at the end of this prospectus, which have been prepared on the same basis as the audited consolidated financial statements. In the opinion of management, the unaudited data reflects all adjustments, consisting only of normal recurring adjustments, necessary for a fair statement of the financial information in those statements. Our historical results are not necessarily indicative of results that may be expected in any future period, and our results for any interim period are not necessarily indicative of results that may be expected for any full year.

	Year ended December 31,		Nine months ended September 30,	
	2016	2017	2017	2018
	(in thousands, except share and per share data)			
Consolidated Statement of Operations Data:				
Operating expenses:				
Research and development	$ 17,439	$ 56,357	$ 26,426	$ 50,475
General and administrative	6,563	9,043	6,263	13,550
Total operating expenses	24,002	65,400	32,689	64,025
Loss from operations	(24,002)	(65,400)	(32,689)	(64,025)
Interest income	65	529	396	2,992
Loss before benefit (provision) for income taxes	(23,937)	(64,871)	(32,293)	(61,033)
Benefit (provision) for income taxes	(36)	(2)	121	386
Net loss	$ (23,973)	$ (64,873)	$ (32,172)	$ (60,647)
Net loss per share attributable to common shareholders—basic and diluted(1)	$ (91.61)	$ (35.85)	$ (19.21)	$ (2.62)
Weighted average common shares outstanding—basic and diluted(1)	261,695	1,809,751	1,675,133	23,174,841

(1) See Note 11 to our annual consolidated financial statements and Note 10 to our interim consolidated financial statements included elsewhere in this prospectus for further details on the calculation of basic and diluted net loss per share attributable to common shareholders.

	As of December 31,		As of September 30,
	2016	2017	2018
			(in thousands)
Consolidated Balance Sheet Data:			
Cash, cash equivalents and short-term investments	$ 55,970	$ 45,555	$ 337,863
Working capital(1)	54,032	29,674	313,507
Total assets	56,467	47,492	347,101
Convertible preferred shares	79,897	119,770	—
Total shareholders' equity (deficit)	(25,732)	(89,708)	318,913

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

Management's discussion and analysis of financial condition and results of operations

You should read the following discussion and analysis of our financial condition and results of operations together with our consolidated financial statements and the related notes and the other financial information included elsewhere in this prospectus. Some of the information contained in this discussion and analysis or set forth elsewhere in this prospectus, including information with respect to our plans and strategy for our business and related financing, includes forward-looking statements that involve risks and uncertainties. As a result of many factors, including those factors set forth in the "Risk factors" section of this prospectus, our actual results could differ materially from the results described in or implied by these forward-looking statements.

Overview

We are a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Our pipeline of five product candidates, across various stages of development, focuses 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-045 and KPL-404.

Our lead candidate is rilonacept, an interleukin-1a, or IL-1a, and interleukin-1b, or IL-1b, cytokine trap. We are developing rilonacept for the potential treatment of recurrent pericarditis, an inflammatory cardiovascular disease and are enrolling a single, pivotal, placebo-controlled, randomized-withdrawal design, global Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis, named RHAPSODY. We also have an ongoing 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 presented interim data from this trial in December 2018 and expect to present additional data at ACC in the first half of 2019.

Mavrilimumab is a monoclonal antibody that antagonizes colony stimulating factor, or GM-CSF. We are evaluating mavrilimumab for the potential treatment of giant cell arteritis, or GCA, an inflammatory disease of the blood vessels. We have commenced dosing in multiple countries in a double-blind, randomized, placebo-controlled, Phase 2 proof-of-concept trial for the study of mavrilimumab in GCA. In the United States, the FDA initially placed our IND for the trial on clinical hold due to its request for additional information regarding the 510(k)-cleared delivery device to be used in our trial. We have since provided the FDA with the requested information and our IND is now active. We plan for U.S. subjects to be included in our ongoing, global Phase 2 proof-of-concept clinical trial.

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 OSMRb. We plan to study KPL-716 in a variety of pruritic, inflammatory, and fibrotic indications driven by these cytokines. In September 2018, we announced interim results from the randomized, double-blind, placebo-controlled, single-ascending-dose, sequential-group portion of the Phase 1a/1b clinical trial in healthy volunteers and in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. We are enrolling a 12-week, repeated single-dose cohort as an additional part of the Phase 1b portion of the Phase 1a/1b clinical trial in subjects with moderate-to-severe atopic dermatitis

experiencing moderate-to-severe pruritus. We expect to report top-line data from this cohort in the second half of 2019. We plan to initiate an adaptive design Phase 2a/2b clinical trial in prurigo nodularis in the first half of 2019 and expect to report top-line data from the first part of this trial in the first half of 2020. We also plan to initiate an exploratory, pilot Phase 2 clinical trial in the first half of 2019 designed to explore the role of IL-31 and OSM in a number of diseases characterized by chronic pruritus and plan to report top-line data from this trial in the second half of 2020.

KPL-045, is a monoclonal antibody inhibitor of the CD30L co-stimulatory molecule. We are continuing our preclinical activities with KPL-045 in inflammatory diseases driven by T-cell-dependent autoantibody generation and dysregulated T_H effector memory responses and expect to file an IND application with the FDA in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020.

KPL-404 is a monoclonal antibody inhibitor of the CD40 co-stimulatory molecule. We are continuing our pre-clinical activities with KPL-404 in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020. We have a license to conduct research and development on KPL-404 from Primatope, the company that owns or controls the intellectual property related to KPL-404. In January 2019, we exercised our exclusive option to acquire all outstanding capital stock of Primatope, which, subject to a supplemental due diligence period, we would expect to close within 60 days of the option exercise. If we do not close the transaction, our license to intellectual property controlled by Primatope to research, develop and manufacture KPL-404 will terminate, in which case we would cease the development of KPL-404.

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

On May 23, 2018, our registration statement on Form S-1 relating to our IPO was declared effective by the SEC. On May 29, 2018, we completed our IPO of 8,477,777 Class A common shares at $18.00 per share for gross proceeds of $152.6 million. 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 of their over-allotment option to purchase additional shares at $18.00 per share, and we issued and sold 1,006,425 Class A common shares 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.

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.

We have incurred significant operating losses since inception. Our ability to generate product revenue sufficient to achieve profitability will depend heavily on the successful development and eventual commercialization of one or more of our current or future product candidates. Our net losses were

$24.0 million and $64.9 million for the years ended December 31, 2016 and 2017, respectively and were $32.2 million and $60.6 million for the nine months ended September 30, 2017 and 2018, respectively. As of December 31, 2017 and September 30, 2018, we had an accumulated deficit of $91.0 million and $151.6 million, respectively. 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 that we did not incur as a private company.

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

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

As of December 31, 2017 and September 30, 2018, we had cash, cash equivalents and short-term investments of $45.6 million and $337.9 million, respectively. 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 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 pre-clinical 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 pre-clinical and clinical drug substance and product for our research and development programs;
- other costs related to acquiring and manufacturing pre-clinical studies and clinical trial materials, including manufacturing validation batches, as well as investigative sites and consultants that conduct our clinical trials, pre-clinical studies and other scientific development services;
- payments made in cash or equity securities under third-party licensing, acquisition and option agreements;
- employee-related expenses, including salaries and benefits, travel and share-based compensation expense for employees engaged in research and development functions;
- costs related to compliance with regulatory requirements; and
- allocated facilities-related costs, depreciation and other expenses, which include rent and utilities.

We recognize external development costs based on an evaluation of the progress to completion of specific tasks using information provided to us by our service providers. This process involves reviewing open contracts and purchase orders, communicating with our personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of actual costs. Nonrefundable advance payments for goods or services to be received in the future for use in research and development activities are recorded as prepaid expenses. Such amounts are recognized as an expense as the goods are delivered or the related services are performed, or until it is no longer expected that the goods will be delivered or the services rendered.

Our direct research and development expenses are tracked on a program-by-program basis for our product candidates and consist primarily of external costs, such as fees paid to outside consultants, CROs, CMOs and research laboratories in connection with our pre-clinical development, process development, manufacturing and clinical development activities. Our direct research and development expenses by program also include fees incurred under license, acquisition and option agreements. We do not allocate employee costs or facility expenses, including depreciation or other indirect costs, to specific programs because these costs are deployed across multiple programs and, as such, are not separately classified. We use internal resources primarily to conduct our research and discovery as well as for managing our pre-clinical development, process development, manufacturing and clinical development activities.

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

	Year ended December 31,		Nine months ended September 30,	
	2016	2017	2017	2018
			(in thousands)	
Rilonacept(1)	$ —	$ 6,301	$ 5,380	$ 6,550
Mavrilimumab(2)	—	18,000	—	6,210
KPL-716(3)	14,870	24,164	15,370	18,670
KPL-045(4)	—	1,654	1,520	3,070
KPL-404(5)	—	549	500	4,090
Unallocated research and development expenses	2,569	5,689	3,656	11,885
Total research and development expenses	$ 17,439	$ 56,357	$ 26,426	$ 50,475

(1) The amount for the year ended December 31, 2017 includes expense of $5.0 million related to an upfront payment under our license agreement with Regeneron. The amount for the nine months ended September 30, 2017 includes expense of $5.0 million related to an upfront payment made under our license agreement with Regeneron.

(2) The amount for the year ended December 31, 2017 consists of expense of $18.0 million related to an upfront payment and an accrued milestone under our license agreement with MedImmune.

(3) The amount for the year ended December 31, 2016 includes expense of $11.5 million related to an upfront payment and $0.5 million related to a technology transfer payment under our asset purchase agreement with Biogen. The amount for the year ended December 31, 2017 and nine months ended September 30, 2017 includes expense of $4.0 million related to a milestone payment under our asset purchase agreement with Biogen associated with the achievement of a specified clinical milestone event.

(4) The amount for the year ended December 31, 2017 includes expense of $1.5 million related to an upfront payment under our license agreement with Novo Nordisk. The amount for the nine months ended September 30, 2018 includes expense of $0.2 million related to a technology transfer payment under our license agreement with Novo Nordisk. The amount for the nine months ended September 30, 2017 includes expense of $1.5 million related to an upfront payment under our license agreement with Novo Nordisk.

(5) The amount for the year ended December 31, 2017 includes expense of $0.5 million related to upfront payments for the initial option period under our stock purchase option agreement with Primatope. The amount for the nine months ended September 30, 2018, includes expense of $0.5 million related to the extension of the option period under our stock purchase option agreement with Primatope. The amount for the nine months ended September 30, 2017 includes expense related to the $0.5 million initial option period payment under our stock purchase option agreement with Primatope.

Research and development activities are central to our business model. Product candidates in later stages of clinical development generally have higher development costs than those in earlier stages of clinical development, primarily due to the increased size and duration of later-stage clinical trials. As a result, we expect that our research and development expenses will increase substantially over the next several years as we complete our ongoing and planned clinical trials for rilonacept, mavrilimumab and KPL-716, as well as conduct other pre-clinical and clinical development including regulatory filings for our other product candidates and our discovery research efforts and 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 option agreements to acquire the rights to our product candidates.

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

- the scope, progress, outcome and costs of our pre-clinical development activities, clinical trials and other research and development activities;

- establishing an appropriate safety and efficacy profile with Investigational New Drug, or 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 U.S. Federal Drug Administration, or 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, 2017, we had state research and development tax credit carryforwards of approximately $0.1 million available to reduce future tax liabilities, which begin to expire in 2031 through 2032.

Results of operations

Comparison of the nine months ended September 30, 2017 and 2018

The following table summarizes our results of operations for the nine months ended September 30, 2017 and 2018:

	Nine months ended September 30,		
	2017	2018	Change
			(in thousands)
Operating expenses:			
Research and development	$ 26,426	$ 50,475	$ 24,049
General and administrative	6,263	13,550	7,287
Total operating expenses	32,689	64,025	31,336
Loss from operations	(32,689)	(64,025)	(31,336)
Interest income	396	2,992	2,596
Loss before benefit for income taxes	(32,293)	(61,033)	(28,740)
Benefit for income taxes	121	386	265
Net loss	$ (32,172)	$ (60,647)	$ (28,475)

Research and development expenses

	Nine months ended September 30,		
	2017	2018	Change
			(in thousands)
Direct research and development expenses by program:			
Rilonacept	$ 5,380	$ 6,550	$ 1,170
Mavrilimumab	—	6,210	6,210
KPL-716	15,370	18,670	3,300
KPL-045	1,520	3,070	1,550
KPL-404	500	4,090	3,590
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	2,932	9,060	6,128
Other	724	2,825	2,101
Total research and development expenses	$ 26,426	$ 50,475	$ 24,049

Research and development expenses were $50.5 million for the nine months ended September 30, 2018, compared to $26.4 million for the nine months ended September 30, 2017. The increase of $24.1 million was primarily due to an increase in external fees related to our development programs, of which there were five in 2018, while in 2017 there were four, as well as an increase of $8.2 million in unallocated research and development expenses.

The direct costs for our rilonacept program were $6.6 million during the nine months ended September 30, 2018, compared to $5.4 million during the nine months ended September 30, 2017, or an increase of $1.2 million. During the nine months ended September 30, 2018, expenses incurred related to our clinical research and development for our open-label Phase 2 proof-of-concept clinical trial and in preparation for our planned Phase 3 clinical trial compared to the nine months ended September 30, 2017, in which expenses incurred were primarily due to the $5.0 million upfront payment made under our license agreement with Regeneron.

The direct costs of $6.2 million for our mavrilimumab program during the nine months ended September 30, 2018 were due to expenses related primarily to preparation for our planned clinical trials, including a Phase 2 trial in giant cell arteritis and manufacturing process development related expenses. We had no direct costs for our mavrilimumab program during the nine months ended September 30, 2017.

The direct costs for our KPL-716 program were $18.7 million during the nine months ended September 30, 2018, compared to $15.4 million during the nine months ended September 30, 2017, or an increase of $3.3 million. During the nine months ended September 30, 2018, expenses incurred related to manufacturing and development costs for our clinical drug supply and our Phase 1a/1b clinical trial, compared to the nine months ended September 30, 2017, in which expenses incurred also included a $4.0 million milestone payment made upon the achievement of a specified clinical milestone event under our agreement with Biogen.

The direct costs for our KPL-045 program were $3.1 million during the nine months ended September 30, 2018, compared to $1.5 million during the nine months ended September 30, 2017, or an increase of $1.6 million. During the nine months ended September 30, 2018, expenses incurred related to clinical research and development, including manufacturing development costs, compared to the nine months ended September 30, 2017, in which expenses incurred related to the $1.5 million upfront payment under our license agreement with Novo Nordisk.

The direct costs for our KPL-404 program were $4.1 million during the nine months ended September 30, 2018, compared to $0.5 million during the nine months ended September 30, 2017, or an increase of $3.6 million. During the nine months ended September 30, 2018, expenses incurred primarily related to clinical research and development, including manufacturing development costs, compared to the nine months ended September 30, 2017, in which expenses incurred related to the $0.5 million initial option period payments under our stock purchase option agreement with Primatope.

Unallocated research and development expenses were $11.9 million for the nine months ended September 30, 2018 compared to $3.7 million for the nine months ended September 30, 2017. The increase of $8.2 million in unallocated research and development expenses was due to an increase of $6.1 million in personnel-related costs, including share-based compensation, and an increase of $2.1 million in other costs, including 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 nine months ended September 30, 2018 and 2017 included share-based compensation of $1.1 million and $0.2 million, respectively.

General and administrative expenses

General and administrative expenses were $13.6 million for the nine months ended September 30, 2018 compared to $6.3 million for the nine months ended September 30, 2017. The increase of $7.3 million was primarily due to increases of $4.3 million in personnel-related costs and $2.5 million in professional fees. The increase in personnel-related costs was due to the hiring of additional personnel in our general and administrative functions, primarily in our corporate departments, including legal, finance and human resources, as we continued to expand our operations to support the organization. Personnel-related costs for the nine months ended September 30, 2018 and 2017 included share-based compensation of $2.0 million and $0.4 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 becoming a public company.

Interest income

Interest income was $3.0 million for the nine months ended September 30, 2018 compared to $0.4 million for the nine months ended September 30, 2017. The increase was due to both higher average invested balances and higher interest rates on U.S. Treasury notes in 2018.

Benefit for income taxes

We recorded an insignificant benefit for income taxes for the nine months ended September 30, 2018 and 2017.

Comparison of the years ended December 31, 2016 and 2017

The following table summarizes our results of operations for the years ended December 31, 2016 and 2017:

	Year ended December 31,		
	2016	**2017**	**Change**
			(in thousands)
Operating expenses:			
Research and development	$ 17,439	$ 56,357	$ 38,918
General and administrative	6,563	9,043	2,480
Total operating expenses	24,002	65,400	41,398
Loss from operations	(24,002)	(65,400)	(41,398)
Interest income	65	529	464
Loss before provision for income taxes	(23,937)	(64,871)	(40,934)
Provision for income taxes	(36)	(2)	34
Net loss	$ (23,973)	$ (64,873)	$ (40,900)

Research and development expenses

	Year ended December 31,		
	2016	2017	Change
			(in thousands)
Direct research and development expenses by program:			
Rilonacept	$ —	$ 6,301	$ 6,301
Mavrilimumab	—	18,000	18,000
KPL-716	14,870	24,164	9,294
KPL-045	—	1,654	1,654
KPL-404	—	549	549
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	1,837	4,576	2,739
Other	732	1,113	381
Total research and development expenses	$ 17,439	$ 56,357	$ 38,918

Research and development expenses were $56.4 million for the year ended December 31, 2017, compared to $17.4 million for the year ended December 31, 2016. The increase of $38.9 million was primarily due to an increase in external fees related to our development programs as well as an increase of $3.1 million in unallocated research and development expenses.

The direct costs of $6.3 million for our rilonacept program during the year ended December 31, 2017 were due to a $5.0 million upfront payment made under our license agreement with Regeneron, as well as $1.3 million of clinical research and development costs associated with the commencement of our open-label Phase 2 proof-of-concept clinical trial. We had no direct costs for our rilonacept program during the year ended December 31, 2016.

The direct costs of $18.0 million for our mavrilimumab program during the year ended December 31, 2017 were due to an $8.0 million upfront payment made under our license agreement with MedImmune as well as an accrued milestone of $10.0 million, as we have determined the payment related to the milestone to be probable. We had no direct costs for our mavrilimumab program during the year ended December 31, 2016.

The direct costs for our KPL-716 program were $24.2 million during the year ended December 31, 2017, compared to $14.9 million during the year ended December 31, 2016. The increase of $9.3 million in direct costs for our KPL-716 program during the year ended December 31, 2017 was primarily due to expenses related to our Phase 1a/1b clinical trial, including a $4.0 million milestone payment made to Biogen upon the achievement of a specified clinical milestone event, as well as expenses related to our LOTUS-PN observational study, manufacturing development costs for clinical drug supply and other research and development studies. During the year ended December 31, 2016, direct costs for our KPL-716 program included expenses of $11.5 million related to an upfront payment and $0.5 million related to a technology transfer payment, each under our agreement with Biogen.

The direct costs of $1.7 million for our KPL-045 program during the year ended December 31, 2017 were primarily due to a $1.5 million upfront payment made under our license agreement with Novo Nordisk. We had no direct costs for our KPL-045 program during the year ended December 31, 2016.

The direct costs of $0.5 million for our KPL-404 program during the year ended December 31, 2017 were due to $0.5 million of upfront payments made in connection with the initial option period under our stock purchase option agreement with Primatope. We had no direct costs for our KPL-404 program during the year ended December 31, 2016.

Unallocated research and development expenses were $5.7 million for the year ended December 31, 2017, compared to $2.6 million for the year ended December 31, 2016. The increase of $3.1 million in unallocated research and development expenses was due to an increase of $2.7 million in personnel-related costs, including share-based compensation, and an increase of $0.4 million in other costs. The increase in personnel-related costs was primarily due to the hiring of additional personnel in our research and development functions, particularly those responsible for coordinating with CMOs on development and manufacturing of drug supply for our product candidates and coordinating with CROs on the conduct and oversight of our Phase 1a/1b clinical trial and LOTUS-PN observational study for our KPL-716 program and our open-label Phase 2 proof-of-concept clinical trial for our rilonacept program. Personnel-related costs for the years ended December 31, 2017 and 2016 included share-based compensation of $0.3 million and $0.1 million, respectively. The increase in other costs was primarily due to a $0.2 million increase in travel expense and a $0.1 million increase in certain allocated facilities-related costs and information technology expenses.

General and administrative expenses

General and administrative expenses were $9.0 million for the year ended December 31, 2017, compared to $6.6 million for the year ended December 31, 2016. The increase of $2.5 million was primarily due to increases of $1.4 million in personnel-related costs and $1.1 million in professional fees. The increase in personnel-related costs was due to the hiring of additional personnel in our general and administrative functions, primarily in our legal and finance departments, as we continued to expand our operations to support the organization. Personnel-related costs for the years ended December 31, 2017 and 2016 included share-based compensation of $0.6 million and $0.3 million, respectively. Professional fees increased due to legal costs incurred in connection with maintaining and registering worldwide patents and costs associated with our ongoing business operations, as well higher accounting, consulting and market research expenses.

Interest income

Interest income was $0.5 million for the year ended December 31, 2017, compared to $0.1 million for the year ended December 31, 2016. The increase was due to both higher average invested cash balances and higher interest rates on U.S. Treasury securities in 2017.

Provision for income taxes

We recorded an insignificant provision for income taxes for the years ended December 31, 2017 and 2016.

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

On May 23, 2018, our registration statement on Form S-1 relating to our IPO was declared effective by the SEC. On May 29, 2018, we completed our IPO of 8,477,777 Class A common shares at $18.00 per share for gross proceeds of $152.6 million. 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 of their over-allotment option to purchase additional shares at $18.00 per share, and we issued and sold 1,006,425 Class A common shares 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.

As of December 31, 2017 and September 30, 2018, we had cash, cash equivalents and short-term investments of $45.6 million and $337.9 million, respectively.

Cash flows

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

	Year ended December 31,		Nine months ended September 30,	
	2016	2017	2017	2018
			(in thousands)	
Net cash used in operating activities	$ (21,867)	$ (50,219)	$ (26,840)	$ (53,374)
Net cash used in investing activities	(3)	(69)	(65)	(268,858)
Net cash provided by financing activities	42,509	39,873	39,873	346,445
Net increase (decrease) in cash and cash equivalents and restricted cash	$ 20,639	$ (10,415)	$ 12,968	$ 24,213

Operating activities

During the nine months ended September 30, 2018, operating activities used $53.4 million of cash, primarily resulting from our net loss of $60.6 million, partially offset by non-cash charges of $1.9 million and net cash provided by changes in our operating assets and liabilities of $5.4 million. Net cash provided by changes in our operating assets and liabilities for the nine months ended September 30, 2018 consisted of a $6.4 million increase in accrued expenses and a $3.0 million increase in accounts payable, offset by a $4.0 million increase in prepaid expenses and other current assets. 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 accounts payable was primarily due to increased operating activities as well as the timing of vendor invoicing and payments. 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.

During the nine months ended September 30, 2017, operating activities used $26.8 million of cash, primarily resulting from our net loss of $32.2 million, partially offset by non-cash charges of $0.4 million and net cash provided by changes in our operating assets and liabilities of $4.9 million.

During the year ended December 31, 2017, operating activities used $50.2 million of cash, primarily resulting from our net loss of $64.9 million, partially offset by non-cash charges of $0.7 million and net cash provided by changes in our operating assets and liabilities of $13.9 million. Net cash provided by changes in our operating assets and liabilities for the year ended December 31, 2017 consisted of a $14.1 million increase in accrued expenses and a $1.0 million increase in accounts payable, both partially offset by a $1.2 million increase in prepaid expenses and other current assets. The increase in accrued expenses was primarily due to an accrued milestone of $10.0 million related to our mavrilimumab

program, increased clinical trial and manufacturing activities as well as increased accrued legal and professional fees and accrued employee compensation-related expenses. The increase in accounts payable was due to our increased level of operating activities and the timing of vendor invoicing and payments. The increase in prepaid expenses and other current assets was primarily due to prepaid clinical trial and manufacturing costs associated with our research and development programs.

During the year ended December 31, 2016, operating activities used $21.9 million of cash, primarily resulting from our net loss of $24.0 million, partially offset by non-cash charges of $0.3 million and net cash provided by changes in our operating assets and liabilities of $1.8 million. Net cash provided by changes in our operating assets and liabilities for the year ended December 31, 2016 consisted primarily of a $1.9 million increase in accrued expenses, partially offset by a $0.2 million increase in prepaid expenses and other current assets. The increase in accrued expenses was primarily due to increased research and development costs, an accrued expense related to technology transfer, regulatory consulting costs and accrued compensation expense. The increase in prepaid expenses and other current assets was primarily due to prepaid manufacturing costs and recording an income tax receivable.

Investing activities

During the nine months ended September 30, 2018, investing activities used $268.9 million of cash, consisting of $1.3 million of purchases of property and equipment and $292.6 million of purchases of short-term investments offset by $25.0 million from proceeds of maturities of short-term investments.

During the nine months ended September 30, 2017, cash used in investing activities was not significant.

During the year ended December 31, 2017, investing activities used $0.1 million of cash, consisting of purchases of property and equipment.

During the year ended December 31, 2016, we used an insignificant amount of cash in investing activities, consisting of purchases of property and equipment.

Financing activities

During the nine months ended September 30, 2018, net cash provided by financing activities was $346.4 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, inclusive of the over-allotment option exercise, and $190.8 million in net proceeds from our issuance and sale of Series C preferred shares, partially offset by $3.6 million of payments of other offering costs associated with our IPO, inclusive of the over-allotment option exercise.

During the nine months ended September 30, 2017, net cash provided by financing activities was $39.9 million, consisting of net proceeds from our issuance and sale of Series B preferred shares.

During the year ended December 31, 2017, net cash provided by financing activities was $39.9 million, consisting of net proceeds from our issuance and sale of Series B preferred shares.

During the year ended December 31, 2016, net cash provided by financing activities was $42.5 million, consisting of net proceeds from our issuance and sale of Series A preferred shares.

Funding requirements

We expect our expenses to increase substantially in connection with our ongoing activities, particularly as we advance the clinical trials and pre-clinical 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 that we did not incur as a private company. Our expenses will also increase as we:

- continue to conduct our current clinical trials and initiate our planned clinical trials, for rilonacept, mavrilimumab and KPL-716;
- advance pre-clinical development of our early-stage programs, KPL-045 and KPL-404;
- manufacture, or have manufactured on our behalf, our pre-clinical and clinical drug material and develop processes for late state and commercial manufacturing;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- establish a sales, marketing, medical affairs and distribution infrastructure to commercialize any product candidates for which we may obtain marketing approval and intend to commercialize on our own;
- hire additional clinical, quality control and scientific personnel;
- expand our operational, financial and management systems and increase personnel, including personnel to support our clinical development, manufacturing and commercialization efforts and our operations as a public company;
- maintain, expand and protect our intellectual property portfolio; and
- in-license or acquire other product candidates and technologies and/or their related businesses, including the payment of up to $18 million in a combination of cash and Kiniksa Class A common shares if we close the acquisition of all of the outstanding capital stock of Primatope.

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 twelve months. We have based these estimates on assumptions that may prove to be wrong, and we could utilize our available capital resources sooner than we expect. We anticipate that we may require additional capital if we choose to pursue in-licenses or acquisitions of other product candidates. If we receive regulatory approval for 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 pre-clinical and clinical trials;
- the costs, timing and outcome of regulatory review of our product candidates;
- the costs, timing and ability to manufacture our product candidates to supply our clinical and pre-clinical development efforts and our clinical trials;
- the costs of future activities, including product sales, medical affairs, marketing, manufacturing and distribution, for any of our product candidates for which we receive marketing approval;

- the costs of manufacturing commercial-grade product and necessary inventory to support commercial launch;
- the ability to receive additional non-dilutive funding, including grants from organizations and foundations;
- the revenue, if any, received from commercial sale of our products, should any of our product candidates receive marketing approval;
- the costs of preparing, filing and prosecuting patent applications, maintaining and enforcing our intellectual property rights and defending intellectual property-related claims;
- our ability to establish and maintain collaborations on favorable terms, if at all;
- the extent to which we acquire or in-license other product candidates and technologies; and
- the timing, receipt and amount of sales of, or milestone payments related to or royalties on, our current or future product candidates, if any.

Until such time, if ever, as we can generate substantial product revenue, we expect to finance our cash needs through a combination of public or private equity offerings, debt financings, governmental funding, collaborations, strategic partnerships or marketing, distribution or licensing arrangements with third parties. To the extent that we raise additional capital through the sale of equity or convertible debt securities, 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

The following table summarizes our contractual obligations as of December 31, 2017 and the effects that such obligations are expected to have on our liquidity and cash flows in future periods:

	Payments due by period				
	Less than 1 year	1 to 3 years	4 to 5 years	More than 5 years	Total
					(in thousands)
Accrued milestone(1)	$ 10,000	$ —	$ —	$ —	$ 10,000
Manufacturing commitments(2)	7,766	—	—	—	7,766
Operating lease commitments(3)	270	—	—	—	270
Total	$ 18,036	$ —	$ —	$ —	$ 18,036

(1) Represents a payment of $10.0 million we are obligated to make under our license agreement with MedImmune upon the earlier to occur of (a) the first achievement of a specified regulatory milestone for a product licensed under the agreement and (b) December 31, 2018.

(2) Amounts in the table reflect commitments for costs associated with our external CMOs, which we have engaged to manufacture pre-clinical and clinical trial materials. Manufacturing commitments include agreements that are enforceable and legally binding on us and that specify all significant terms, including fixed or minimum quantities to be purchased; fixed, minimum or variable price provisions; and the approximate timing of the transaction. For obligations with cancellation provisions, the amounts included in the preceding table are limited to the non-cancelable portion of the agreement terms or the minimum cancellation fee. As of September 30, 2018, non-cancelable commitments associated with our external CMOs were $34.9 million.

(3) Represents minimum payments due for the lease of office space by our wholly owned U.S. subsidiary, Kiniksa Pharmaceuticals Corp., or Kiniksa US, in Wellesley Hills, Massachusetts under an operating lease agreement that expired in August 2018. In March 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 in June 2018, Kiniksa US entered into an amendment to the lease expanding the rentable space. In November 2018, Kiniksa US entered into an amendment to the lease expanding the rentable space, which will be occupied in phases through December 2019. The lease expires in July 2021. Monthly lease payments include base rent as well as, ancillary charges such as the share of operating expenses and real estate taxes. Base rent is approximately $0.1 million per month. The lease requires future rental payments of $0.2 million during the year ending December 31, 2018, an aggregate of $2.9 million during the years ending December 31, 2019 and 2020 and $1.0 million during the year ending December 31, 2021. Such amounts are not reflected in the table.

Our contracts with CMOs, CROs and other third parties for the manufacture of our product candidates and to support clinical trials and pre-clinical research studies and testing are generally cancelable by us upon prior notice. Payments due upon cancellation consisting only of payments for services provided or expenses incurred, including noncancelable obligations of our service providers, up to the date of cancellation are not included in the preceding table as the amount and timing of such payments are not known.

Under various agreements with third parties, we have agreed to make milestone payments, pay royalties, annual maintenance fees and to meet due diligence requirements based upon specified milestones. We generally have not included any contingent payment obligations, such as milestones, royalties or due diligence, in the table above as the amount, timing and likelihood of such payments are not known. We have not included any of the annual maintenance fee payments in the above table, as although the amount and timing are known, we cannot currently determine the final termination dates of the agreements and, as a result, we cannot determine the total amounts of such payments we will be required to make under the agreements.

Under our license agreement with Regeneron, we are obligated to make future regulatory milestone payments of $27.5 million in the aggregate. Thereafter, we have agreed to evenly split profits on our sales of rilonacept with Regeneron after deducting certain commercialization expenses subject to specified limits.

Under our license agreement with MedImmune, we are obligated to make future clinical, regulatory and initial sales milestone payments of up to $72.5 million in aggregate for the first two indications we develop, including, a $5.0 million pass-through payment due upon the achievement of a specified clinical milestone event which is anticipated to be met in the fourth quarter of 2018. Also included is a milestone payment of $10.0 million upon the earlier to occur of a specified regulatory milestone and December 31, 2018, and clinical and regulatory milestone payments of up to $15.0 million in the aggregate for each subsequent indication. The $10.0 million milestone payment was accrued on our consolidated balance sheet as of December 31, 2017 and recognized as research and development expense during the year ended December 31, 2017. Such payment is included in the table above. We are also obligated to make milestone payments to MedImmune of up to $85.0 million upon the achievement of annual net sales thresholds of up to, but excluding, $1.0 billion in annual net sales as well as additional milestone payments aggregating up to $1.1 billion upon the achievement of additional specified annual net sales thresholds starting at $1.0 billion and higher. Commencing on the first commercial sale of licensed products, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. We must pay such royalties on a product-by-product and country-by-country basis until the latest to occur of the expiration of licensed patents, the expiration of regulatory exclusivity or the tenth anniversary of first commercial sale of such product in such country.

Under our asset purchase agreement with Biogen, we are obligated to make future milestone payments of up to $325.0 million upon the achievement of specified clinical and regulatory milestones as well as upon the achievement of annual net sales thresholds. We have also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to KPL-716. Additionally, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

Under our license agreement with Novo Nordisk, we are obligated to make future milestone payments upon the achievement of specified clinical, regulatory, initial sales milestones as well as upon the achievement of annual net sales thresholds, including a payment of $1.0 million upon the earlier to occur of a specified regulatory milestone and January 2020. We are also obligated to pay royalties on annual net sales of products licensed under the agreement. In addition, we are obligated to make a payment upon the completion of technology transfer.

In January 2019, we exercised our exclusive option to acquire all outstanding capital stock of Primatope, which, subject to a supplemental due diligence period, we would expect to close within 60 days of the option exercise. If we close the transaction, we will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10.0 million as well as potential milestone payments of up to $8.0 million ($3.0 million of which has been achieved as of the date of this prospectus and would be payable at closing), payable in a combination of cash and our Class A common shares.

Critical accounting policies and significant judgments and estimates

Our consolidated financial statements are prepared in accordance with generally accepted accounting principles in the United States. The preparation of our consolidated financial statements and related disclosures requires us to make estimates and judgments that affect the reported amounts of assets, liabilities, costs and expenses, and the disclosure of contingent assets and liabilities in our financial statements. We base our estimates on historical experience, known trends and events and various other factors that we believe are reasonable under the circumstances, the results of which form the basis for making judgments about the carrying values of assets and liabilities that are not readily apparent from other sources. We evaluate our estimates and assumptions on an ongoing basis. Our actual results may differ from these estimates under different assumptions or conditions.

While our significant accounting policies are described in more detail in Note 2 to our consolidated financial statements appearing at the end of this prospectus, we believe that the following accounting policies are those most critical to the judgments and estimates used in the preparation of our consolidated financial statements.

Accrued research and development expenses

As part of the process of preparing our consolidated financial statements, we are required to estimate our accrued research and development expenses. This process involves reviewing open contracts and purchase orders, communicating with our personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of actual costs. The majority of our service providers invoice us in arrears for services performed, on a pre-determined schedule or when contractual milestones are met; however, some require advanced payments. We make estimates of our accrued expenses as of each balance sheet date in the consolidated financial statements based on facts and circumstances known to us at that time. We periodically confirm the accuracy of these estimates with the service providers and make

adjustments if necessary. Examples of estimated accrued research and development expenses include fees paid to:

- vendors, including research laboratories, in connection with pre-clinical development activities;
- CROs and investigative sites in connection with pre-clinical studies and clinical trials; and
- CMOs in connection with drug substance and drug product formulation of pre-clinical studies and clinical trial materials.

We base our expenses related to pre-clinical studies and clinical trials on our estimates of the services received and efforts expended pursuant to quotes and contracts with multiple research institutions and CROs that conduct and manage pre-clinical studies and clinical trials on our behalf. The financial terms of these agreements are subject to negotiation, vary from contract to contract and may result in uneven payment flows. There may be instances in which payments made to our vendors will exceed the level of services provided and result in a prepayment of the expense. Payments under some of these contracts depend on factors such as the successful enrollment of patients and the completion of clinical trial milestones. In accruing service fees, we estimate the time period over which services will be performed and the level of effort to be expended in each period. If the actual timing of the performance of services or the level of effort varies from the estimate, we adjust the accrual or the amount of prepaid expenses accordingly. Although we do not expect our estimates to be materially different from amounts actually incurred, our understanding of the status and timing of services performed relative to the actual status and timing of services performed may vary and may result in reporting amounts that are too high or too low in any particular period. To date, there have not been any material adjustments to our prior estimates of accrued research and development expenses.

Share-based compensation

We measure options and other share-based awards granted to employees and directors based on the fair value on the date of the grant and recognize the corresponding compensation expense of those awards over the requisite service period, which is generally the vesting period of the respective award. We have only issued options and restricted share awards with service-based vesting conditions and record the expense for these awards using the straight-line method.

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

We estimate the fair value of each option grant using the Black-Scholes option-pricing model, which uses as inputs the fair value of our Class A common shares and assumptions we make for the volatility of our Class A common shares, the expected term of our options, the risk-free interest rate for a period that approximates the expected term of our options and our expected dividend yield. Until our IPO, we were a private company and we lacked company-specific historical and implied volatility information. Accordingly, we estimate our expected volatility based on the historical volatility of publicly traded peer companies and expect to continue to do so until such time as we have adequate historical data regarding the volatility of our traded share price. The expected term of our options has been determined utilizing the "simplified" method for awards that qualify as "plain-vanilla" options, while the expected term of our options granted to consultants and nonemployees has been determined based on the contractual term of the options. The

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

Emerging growth company status

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

Off-balance sheet arrangements

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

Recently issued accounting pronouncements

A description of recently issued accounting pronouncements that may potentially impact our financial position and results of operations is disclosed in Note 2 to our annual consolidated financial statements and our interim consolidated financial statements appearing at the end of this prospectus.

Quantitative and qualitative disclosures about market risks

Interest rate risk

We are exposed to market risk related to changes in interest rates. As of December 31, 2016, December 31, 2017 and September 30, 2018, 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.

Business

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 five product candidates, across various stages of development, focused on autoinflammatory and autoimmune conditions. We have three clinical-stage product candidates and two pre-clinical-stage product candidates. We follow a disciplined and methodical approach to selectively identify, discover and acquire product candidates with strong biologic rationales or validated mechanisms of action. We believe that each of our product candidates has the potential to address multiple indications.

Our portfolio of product candidates offers multiple development opportunities. By modulating different parts of the innate and adaptive immune system, these product candidates together have the potential to provide a variety of mechanisms to address multiple devastating diseases.

- ***Rilonacept*** is a protein cytokine trap for inhibiting interleukin-1a, or IL-1a, and interleukin-1b, or IL-1b. Cytokines are small proteins that play a key role in cell signaling. Rilonacept is approved by the U.S. Food and Drug Administration, or the FDA, for the treatment of Cryopyrin-Associated Periodic Syndromes, or CAPS, and has been commercially sold as ARCALYST by Regeneron Pharmaceuticals, Inc., or Regeneron, for this indication since 2008. We licensed rilonacept from Regeneron in 2017. We are initially developing rilonacept for the treatment of recurrent pericarditis, a debilitating inflammatory cardiovascular disease. We are not aware of any therapy currently approved by the FDA for the treatment of recurrent pericarditis. We are enrolling a single, pivotal, global, Phase 3 clinical trial in recurrent pericarditis, named RHAPSODY. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data in the second half of 2020. RHAPSODY is a double-blind, placebo-controlled, randomized-withdrawal, or RW, design study with open-label extension which is designed to assess the efficacy and safety of rilonacept treatment in subjects with recurrent pericarditis. We also have an ongoing open-label Phase 2 proof-of-concept clinical trial in recurrent pericarditis, for which we have recently completed enrollment. In December 2018, we reported interim data from the Phase 2 trial. As of the November 1, 2018 data cutoff date, interim data from 12 symptomatic subjects participating in one portion of the phase 2 trial showed a reduction in both c-reactive protein, or CRP, an inflammation biomarker, and reported pain. As of the cutoff date, 10 of the subjects had completed the 6-week base treatment period and entered into the optional 18-week extension period. Four of the 10 subjects had completed the optional 18-week extension period. All subjects showed a persistent clinical response as measured by c-reactive protein and pain levels at each measurement point during the study. Rilonacept has been generally well-tolerated in the trial, with adverse events, or AEs, consistent with the FDA-approved label for the treatment of CAPS. The most common AEs were gastrointestinal disorders and injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment. Infections are reported in the rilonacept label. We expect to present additional data at the American College of Cardiology 68th Annual Scientific Session & EXPO 2019, or ACC, in the first half of 2019.

- ***Mavrilimumab*** is a monoclonal antibody that antagonizes the signaling of granulocyte macrophage colony stimulating factor, or GM-CSF. We are focusing our initial development efforts for mavrilimumab on giant cell arteritis, or GCA, an inflammatory disease of the blood vessels with unmet medical need that can lead to blindness if left untreated. MedImmune Limited, or MedImmune, initially developed mavrilimumab for the treatment of rheumatoid arthritis, or RA. MedImmune's Investigational New Drug

application, or IND, for the clinical development of mavrilimumab for the treatment of RA was initially put on clinical hold in 2010 before human data had been generated due to certain effects that were observed in non-clinical studies, which coincides with a theoretical risk of developing pulmonary alveolar proteinosis, or PAP, possibly in the setting of GM-CSF inhibition. Since then, in 2014, the FDA acknowledged that clinical studies in refractory RA may be appropriate based on MedImmune's clinical studies in Europe in which it dosed over 550 RA patients with mavrilimumab with no evidence of PAP. Medimmune has since withdrawn the IND for Mavrilimumab for the treatment of RA, and we submitted a new IND with the FDA for the study of mavrilimumab in GCA. The FDA initially placed our IND on clinical hold due to its request for additional information regarding the 510(k)-cleared delivery device to be used in our Phase 2 clinical trial. We have since provided the FDA with the requested information and our IND is now active. We plan for U.S. subjects to be included in our ongoing, double-blind, randomized, placebo-controlled, global Phase 2 proof-of-concept clinical trial, for which we have commenced dosing in multiple countries. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data 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 OSMRb. We plan to study KPL-716 in a variety of pruritic, inflammatory, and fibrotic indications driven by these cytokines, and we believe KPL-716 is the only monoclonal antibody in development that simultaneously targets both pathways. At the European Association of Dermatology and Venereology congress in September 2018, we presented results from the randomized, double-blind, placebo-controlled, single-ascending-dose, sequential-group portion of the Phase 1a/1b clinical trial in healthy volunteers and in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. In this trial, single intravenous, or IV, and subcutaneous, or SC, doses were well tolerated. The results provided an early signal in efficacy in reducing pruritus (assessed by the Worst-Itch Numerical Rating Scale), as well as reducing inflammation and disease severity (assessed by Eczema Area Severity Index, or EASI) in atopic dermatitis subjects after a single dose of KPL-716 in a placebo-controlled, exploratory efficacy assessment (20 subjects were randomized 1:1 and received either 7.5 mg/kg IV of KPL-716 or placebo). To help us to understand whether KPL-716 could be a competitive therapeutic in atopic dermatitis, if approved, we are enrolling a 12-week, repeated single-dose cohort as an additional part of the Phase 1b portion of the Phase 1a/1b clinical trial in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. This cohort is designed to evaluate safety, tolerability, pharmacokinetics and immunogenicity, and it will allow us to conduct an exploratory efficacy analysis on both pruritus as well as disease severity response markers (assessed by EASI). We expect to report top-line data from this cohort in the second half of 2019. We also believe that the single-dose results from the Phase 1a/1b study provide proof-of-principle for KPL-716's potential to treat a spectrum of diseases that may have pruritus mediated by IL-31 and support our plans to advance KPL-716 into multiple diseases where chronic pruritus plays a role in patient symptomatology. We plan to initiate an adaptive design Phase 2a/2b clinical trial in prurigo nodularis in the first half of 2019 and expect to report top-line data from the first part of this trial in the first half of 2020. We also plan to initiate an exploratory, pilot Phase 2 clinical trial in the first half of 2019 designed to explore the role of IL-31 and OSM in a number of diseases characterized by chronic pruritus and to report top-line data from this trial in the second half of 2020.

- ***KPL-045*** is a monoclonal antibody inhibitor of the CD30/CD30L interaction, a T-cell co-stimulatory receptor involved in activated T-memory cell function. We are continuing our pre-clinical activities in KPL-045 in inflammatory diseases driven by T-cell-dependent autoantibody generation and dysregulated

T_H effector memory responses and expect to file an IND with the FDA in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020.

- ***KPL-404*** is a monoclonal antibody inhibitor of the CD40/CD40L interaction, a central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity. We are continuing our pre-clinical activities in KPL-404 in T-cell dependent, B-cell mediated diseases, and expect to file an IND with the FDA for this program in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020. In January 2019, we exercised our exclusive option to acquire all of the outstanding capital stock of Primatope Therapeutics, Inc., or Primatope, the company that owns or controls the intellectual property related to KPL-404. We expect to close this transaction within 60 days of the option exercise subject to a supplemental due diligence period. If we do not close this transaction, our license to the intellectual property controlled by Primatope to research, develop and manufacture KPL-404 will terminate and we will cease the development of KPL-404.

The following table summarizes our current pipeline of product candidates:

Program & Target	Indication	Phase	Status	Rights
Rilonacept[1] IL-1α & IL-1β	Recurrent Pericarditis (RP)	Preclin 1 2 3	• Enrolling single, pivotal Phase 3 trial in recurrent pericarditis; top-line data expected in 2H 2020 • Phase 2 trial data presentation at ACC expected in 1H 2019	Worldwide (excluding MENA)
Mavrilimumab GM-CSFRα	Giant Cell Arteritis (GCA)		• Enrolling global Phase 2 proof-of-concept trial • Top-line data expected in 2H 2020	Worldwide
KPL-716 OSMRβ	Prurigo Nodularis (PN)		• Plan to initiate adaptive design Phase 2a/2b trial in PN in 1H 2019 • Top-line data from Phase 2a expected in 1H 2020	Worldwide
	Chronic Idiopathic Urticaria, Chronic Idiopathic Pruritus, Lichen Planus, Lichen Simplex Chronicus & Plaque Psoriasis		• Plan to initiate Phase 2 exploratory pilot trial in multiple diseases characterized by chronic pruritus in 1H 2019 • Top-line data from Phase 2 trial expected in 2H 2020	
	Atopic Dermatitis (AtD)		• Top-line data from repeated-single-dose Phase 1b trial expected in 2H 2019	
KPL-045[2] CD30L	Autoimmune		• IND filing planned for 2H 2019 • Plan to initiate Phase 1 trial in 1H 2020	Worldwide
KPL-404[2 3] CD40	Autoimmune		• IND filing planned for 2H 2019 • Plan to initiate Phase 1 trial in 1H 2020	Worldwide

(1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication.

(2) We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.

(3) Subject to closing the acquisition of Primatope.

In addition to the indications described above, we plan to evaluate rilonacept, mavrilimumab and KPL-716 in other indications. We plan to be opportunistic in our business development activities to identify and potentially acquire the rights to additional programs or companies that could expand our existing portfolio and/or platform capability. We have also initiated our own internal research efforts to discover and develop molecules to address areas of unmet medical need.

We currently plan to commercialize our product candidates, if approved, in the United States and select international markets. In parallel with our product development timelines, we plan to build our own commercial and operational organizations around the world as appropriate. We anticipate building targeted

medical affairs and sales teams focused on specialist physicians who treat the patient populations addressed by our product candidates.

Our team

We have assembled an experienced management team with a successful track record. Our team has expertise across the spectrum of global drug discovery, development, manufacturing and commercialization activities in diseases within both large and orphan indications. Our Chairman and Chief Executive Officer, Sanj K. Patel, has more than 25 years of scientific, clinical and commercial experience in the pharmaceutical and biotechnology industries. Our Chief Medical Officer, John F. Paolini, M.D., Ph.D., has more than 17 years of experience planning, operating and executing clinical development programs across a range of disease indications from orphan diseases to large cardiovascular diseases, and ten years as a practicing cardiologist. Other members of our senior management team have held key management positions at other companies that developed and commercialized therapies for underserved, rare and specialty-focused patient populations. These companies include Synageva, Genzyme, Novo Nordisk, Shire, Sanofi, Pfizer, Bayer, Merck, Novartis and Vertex, among others.

Our strategy

Our vision is to build a fully-integrated, global biopharmaceutical company by discovering, acquiring, developing and commercializing life-changing therapies for debilitating diseases. We are developing a pipeline of novel drug product candidates for the treatment of autoinflammatory and autoimmune diseases, and we aim to be an industry leader in these areas. We are pursuing multiple programs in parallel, with the goal of delivering safe and effective therapies to patients as efficiently as possible.

Critical components of our business strategy include the following:

- **Efficiently and rapidly advance our product candidates through the development process.** We believe that our product candidates have the potential to address significant unmet medical needs and intend to develop them as efficiently and rapidly as possible.

- **Commercialize our product candidates to bring new or improved therapies to patients in need.** We intend to market and commercialize our product candidates, if approved, in the United States and select international markets by developing our own sales, marketing, medical affairs and reimbursement organizations as appropriate. We anticipate creating a targeted sales organization that supports specialist physicians who treat these specific patient populations and plan to build out this organization as our product candidates approach potential regulatory approval. We believe this approach will allow us to effectively reach patients and prescribers that our product candidates target and leverage the commercial potential of our product candidates.

- **Maximize our existing portfolio opportunity by expanding use across multiple indications.** A core component of our approach to product development is identifying assets that each have the potential to treat multiple diseases. We aim to develop and commercialize our product candidates to produce meaningful impact for patients across relevant indications. Our assets are designed to modulate signaling pathways that are implicated across a spectrum of autoimmune and autoinflammatory conditions. We believe that all of our product candidates have potential in multiple indications.

- **Leverage our value-driven approach to identify, discover, acquire and develop new therapies.** We follow a disciplined and methodical approach to our review of new opportunities. We focus on research-based and comprehensive indication mapping exercises to categorize and prioritize indications of

interest. We evaluate a variety of factors for potential product candidates, technologies and discovery targets, including biologic rationale for addressing the disease, potential for regulatory approval, commercial viability, intellectual property position, prospects for favorable pricing and reimbursement and the impact of competition. We also look at assets that could potentially address multiple indications. In building our current pipeline, we evaluated a large number of opportunities and negotiated agreements with parties for the assets that met our criteria and have in-licensed or acquired the rights to develop and commercialize five separate biologics. Going forward, we intend to be opportunistic in our business development activities.

- **Build our core capability in autoimmune and autoinflammatory diseases to establish a leadership position in the field.** Our pipeline consists of protein therapeutic candidates across various stages of drug development, including a cytokine trap—rilonacept—and four monoclonal antibodies—mavrilimumab, KPL-716, KPL-045 and KPL-404. Both categories of therapeutics functionally inhibit signaling pathways that are implicated in autoinflammatory- or autoimmune-driven pathologies. We intend to leverage our internal discovery efforts and business development capabilities to complement our existing portfolio to build our core capability and establish a leadership position in the field.

Our product candidates

Rilonacept

Overview

Rilonacept was approved by the FDA for the treatment of cryopyrin-associated periodic syndromes, or CAPS, which includes cold auto-inflammatory syndrome and Muckle-Wells syndrome, and has been commercially sold as ARCALYST in the United States by Regeneron for this indication since 2008. We licensed rilonacept in 2017 from Regeneron. We believe that rilonacept has potential to treat certain diseases mediated by both IL-1a and IL-1b. Our lead indication for rilonacept is recurrent pericarditis, which is a recurring painful inflammation of the pericardium. We are enrolling a single, pivotal, global, Phase 3 clinical trial in recurrent pericarditis, named RHAPSODY. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data in the second half of 2020. We also have an ongoing open-label Phase 2 proof-of-concept clinical trial in order to gain further experience with rilonacept in different pericarditis populations and reported interim results of this trial in December 2018. We expect to present additional data from this trial at ACC in the first half of 2019. We plan to evaluate rilonacept in additional diseases mediated by IL-1a and IL-1b.

There is currently one other FDA-approved agent that blocks both IL-1a and IL-1b signaling, anakinra, and one that blocks only IL-1b, canakinumab. We believe both therapies have limitations, and neither is approved by the FDA for treatment of pericarditis. Anakinra requires once-daily injections, and canakinumab only blocks IL-1b, making it less effective or ineffective in diseases driven by IL-1a pathology. We believe that rilonacept with its more moderate, once-weekly dosing schedule and its ability to inhibit both IL-1a and IL-1b could provide an improved therapeutic option for a variety of IL-1a-mediated diseases.

Mechanism of action

Rilonacept is an inhibitor of IL-1a and IL-1b. IL-1a and IL-1b have been demonstrated to play a key role in inflammatory diseases. IL-1a and IL-1b provoke potent, pro-inflammatory events by engaging the IL-1a and IL-1b receptor. Following tissue insult, the release of IL-1a acts as the primary initiating signal to coordinate the mobilization of immune cells to the damaged area, while IL-1b is secreted mostly by macrophages and is a prototypical cytokine of the canonical inflammasome. IL-1a and IL-1b signaling

results in a dramatic increase in the production of cytokines that orchestrate the proliferation and recruitment of phagocytes to the site of damage, resulting in inflammation. Moreover, IL-1a and IL-1b signaling also affect other immune-system cells, such as T-cells and B-cells.

IL-1b's role in the inflammation process has been extensively studied, while in comparison, much is still unknown about the independent function of IL-1a in disease pathology. Despite driving similar immunological outcomes, IL-1a and IL-1b differ substantially in their expression and regulation, and non-redundant roles for IL-1a or IL-1b have been demonstrated in multiple inflammatory diseases. There are disease states in which IL-1b inhibition alone does not appear to be sufficient for disease remission in the absence of IL-1a inhibition. Published studies suggest certain autoinflammatory diseases may, in fact, be pathologically driven primarily by IL-1a.

An investigator-initiated study of anakinra successfully demonstrated mechanistic proof-of-concept for inhibiting both IL-1a and IL-1b in the treatment of recurrent pericarditis. In a published case study, a patient with a refractory form of recurrent pericarditis, who was well-controlled on anakinra, was switched from anakinra to canakinumab, which inhibits only IL-1b, for tolerability reasons. The patient's disease returned despite further dose escalation of canakinumab. When the patient was switched back to anakinra, which inhibits IL-a and IL-b, the disease promptly went back into remission. These data, together with clinical data from our ongoing open-label Phase 2 proof-of-concept study and confirmatory market research, may indicate that IL-1a and IL-1b play unique roles in recurrent pericarditis and other autoinflammatory diseases in which the pathology may be driven primarily by IL-1a.

Background and market opportunity for recurrent pericarditis

Pericarditis is the most common disorder involving the pericardium, the two-layered sac that surrounds the heart. Pericarditis is an inflammation of this sac and is typically characterized by significant chest pain, shortness of breath, coughing and fatigue and is often misconstrued by patients as a heart attack. In addition, typical signs of pericarditis include pericardial friction rub, electrocardiogram changes or pericardial effusion, which is a build-up of fluid around the heart. Pericarditis is described as recurrent if, following an initial occurrence of pericarditis, it recurs after a symptom-free period of about four to six weeks. Pericarditis is considered chronic if symptoms of any one episode last longer than three months, typically causing significant pain and frustration. If pericarditis is left untreated, patients can develop thickening and scarring of the pericardium, potentially requiring invasive surgical stripping. Pericardial effusion, if large enough, can compress the heart externally, requiring emergent drainage.

We intend to focus our development of rilonacept for the treatment of recurrent pericarditis initially in the United States, and we are exploring opportunities for potential expansion into other countries. Claims analysis, cross validated with published estimates, supports a prevalent population of patients with recurrent pericarditis seeking and receiving medical treatment to be approximately 40,000. Within this estimated diagnosed and treated recurrent pericarditis patient population, there are certain subgroups of patients totaling approximately 14,000 with particularly high unmet medical needs consisting of:

- patients who are refractory to conventional treatments (approximately 3,000);
- patients who are dependent on steroids or not well-controlled on their existing therapy (approximately 6,000); and
- patients who are steroid intolerant and refractory to NSAIDs and colchicine (approximately 5,000).

There may be other thoracic inflammatory syndromes where rilonacept may prove beneficial, such as pericarditis associated with post-pericardiotomy syndrome, an inflammatory reaction of the pericardium in

patients who have undergone surgery that involves opening the pericardium. Post-pericardiotomy syndrome occurs in up to 30% of the 300,000 patients in the United States undergoing open heart surgery, and we believe rilonacept may be a therapeutic option for a subset of these patients.

Current treatment landscape for recurrent pericarditis

We are not aware of any current therapies approved by the FDA for the treatment of recurrent pericarditis. A patient's initial acute episode of pericarditis is typically treated with over-the-counter or prescription NSAIDs or colchicine, both of which are used off-label. Recurrent episodes are treated in a similar manner or by adding systemic corticosteroids which are also used off-label. Both colchicine and corticosteroids often have deleterious effects when used at high doses or for long periods of time, including, for colchicine, gastrointestinal distress and neutropenia and, for corticosteroids, glaucoma, fluid retention, hypertension, mood changes, memory changes, other psychological effects, weight gain and diabetes. Fourth-line treatment for these patients may include other immunosuppressants such as methotrexate and azathioprine, as well as anakinra.

Our solution

Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1a and IL-1b signaling. Beyond recurrent pericarditis, we believe there is significant potential for rilonacept to address additional indications, including other pericarditis populations. More broadly, we believe diseases characterized by painful serosal inflammation may be driven by IL-1a, and we intend to consider development of rilonacept in these indications and in others where we believe IL-1a or IL-1b play a key role in disease pathophysiology.

Clinical development plan for recurrent pericarditis

We are enrolling a pivotal, global Phase 3 clinical trial, named RHAPSODY. The study is intended to evaluate the efficacy and safety of rilonacept treatment in subjects with recurrent pericarditis. RHAPSODY is a double-blind, placebo-controlled, RW-study with an open-label extension period. We expect that up to approximately 50 subjects will be randomized into the RW period. Eligible subjects must present at screening with at least a third pericarditis episode, defined as at least 1 day with pericarditis pain ³ 4 on the 11-point NRS and a C-reactive protein, or CRP, value ³1 mg/dL within the 7-day period prior to first study drug administration. Subjects included in the study may be receiving concomitant nonsteroidal anti-inflammatory drugs (NSAIDs) and/or colchicine and/or oral corticosteroid treatment in any combination.

The clinical study is comprised of 5 periods:

- a screening period;
- a single-blind run-in period during which subjects receive a 320 mg loading dose of rilonacept subcutaneously followed by 160 mg SC weekly while background pericarditis medications are tapered and discontinued;
- a double-blind, placebo-controlled 24-week RW period where clinical responders to rilonacept are randomized 1:1 to 160 mg SC weekly rilonacept or placebo;
- a long-term extension treatment period for where all subjects completing the RW period have the option to receive up to 24-weeks of open-label rilonacept 160 mg SC weekly; and

- a long-term extension follow-up period during which all subjects in the long-term extension period will be followed for 24 weeks for safety and pericarditis recurrences.

The primary efficacy endpoint is time-to-first-pericarditis-recurrence in the RW period. The Clinical Endpoint Committee will adjudicate all suspected pericarditis recurrences for inclusion in the primary efficacy endpoint analysis. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data in the second half of 2020.

We have an ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept in recurrent pericarditis in order to gain further experience with rilonacept in different pericarditis populations. This trial evaluates the treatment response to rilonacept 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. The trial is divided into five parts, each enrolling subjects who are currently on any combination of co-administered NSAIDs, colchicine or corticosteroids. The subjects are dosed using the approved rilonacept dose for CAPS, which is a loading dose of two 160 mg subcutaneous doses (320 mg total), followed by single, self-administered 160 mg subcutaneous doses every seven days for a total of six weeks. This is followed by an 18-week extension period. During the extension period, the investigator may choose to wean concomitant NSAIDs, colchicine or corticosteroids according to standard-of-care paradigms. The assessed efficacy outcomes measures include an 11-point pain Numerical Rating Scale or, NRS, CRP, electrocardiogram, and size of pericardial effusion.

The five parts of the trial with different patient populations are:

- Part 1: Symptomatic subjects with recurrent pericarditis receiving NSAIDS +/– colchicine +/– steroids with high CRP, a marker of inflammation, measurements;
- Part 2: Symptomatic subjects with recurrent pericarditis receiving NSAIDS +/– colchicine +/– steroids without elevated CRP measurements but with evidence of pericardial inflammation by MRI;
- Part 3: Subjects with recurrent pericarditis who are dependent upon or unable to wean off of corticosteroids;
- Part 4: Symptomatic subjects with postpericardiotomy syndrome receiving NSAIDS +/– colchicine +/– steroids with high CRP measurements; and
- Part 5: Subjects with postpericardiotomy syndrome who are dependent upon or unable to wean off of corticosteroids.

In December 2018, we reported interim data from Part 1 of the open-label Phase 2 proof-of-concept clinical trial that showed a reduction in both inflammation and reported pain in the 6-week base treatment period and a persistent clinical response during the optional 18-week extension period.

As of the November 1, 2018 interim data analysis cutoff, 12 subjects, each with at least 3 episodes of pericarditis and elevated CRP (>1mg/dL), enrolled in a 6-week base treatment period. Results showed a reduction in both a biomarker of inflammation (CRP) and reported pain (NRS) after the first dose and a persistent clinical response throughout the 6-week base treatment period:

- mean patient-reported pericardial pain on a 11-point NRS decreased from 4.6 at baseline (n=12) to 0.9 at 6 weeks (n=8);

- mean CRP decreased from 4.9 mg/dL at baseline (n=12) to 0.37 mg/dL at 6 weeks (n=4); median time to CRP normalization was 9 days (n=12); and
- pericardial signs resolved, including pericardial effusion (5/6 subjects), PR depression (3/4 subjects), widespread ST elevation (2/2 subjects), and pericardial rub (3/3 subjects).

As of November 1, 2018, 10 of the 12 enrolled subjects received at least 6 weeks of treatment with rilonacept, 6 continued into the optional 18-week extension period, and 4 completed 24 weeks of treatment. These subjects exhibited a continued clinical response to rilonacept, including as described below:

- mean patient-reported pericardial pain on a 11-point NRS was 0.3, and mean CRP was 0.44 mg/dL at 24 weeks (n=4);
- the pericardial effusion in the 1 remaining subject resolved during the extension period; and
- of the 4 subjects on corticosteroids at baseline, the 1 subject who had completed 24 weeks of treatment successfully tapered off corticosteroids.

Rilonacept has been generally well-tolerated in the study, with AEs consistent with the FDA-approved label for the treatment of CAPS, including Familial Cold Auto-inflammatory Syndrome and Muckle-Wells Syndrome. The most common AEs were gastrointestinal disorders and injection site reactions. Seven of 12 subjects experienced at least one treatment-related adverse event during the treatment period. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment. Infections are reported in the rilonacept label for CAPs.

We closed enrollment for the open-label Phase 2 proof-of-concept study in December 2018 and we expect to present additional data from this trial at ACC in the first half of 2019.

Clinical history of rilonacept

Regeneron evaluated rilonacept in a total of 21 clinical trials, including two trials in over 100 patients for the treatment of CAPS, and six trials in over 1,800 patients for the treatment of gout flares.

- *CAPS:* Regeneron evaluated rilonacept for the treatment of CAPS in two trials. In these trials, 109 patients with CAPS, including eight pediatric patients, were treated with at least one dose of rilonacept. In the pivotal efficacy trial, which evaluated the long-term efficacy and safety of once-weekly dosing, 160 mg of rilonacept markedly decreased the clinical signs and symptoms of CAPS.
- *Gout:* Regeneron evaluated rilonacept for the treatment of gout flares in six trials. In the two pivotal efficacy trials in patients with gout, which evaluated the efficacy of once-weekly dosing for the prevention of gout flares during initiation of uric acid-lowering therapy, rilonacept at doses of 80 mg and 160 mg significantly decreased the number of gout flares. Regeneron abandoned active development for the treatment of gout flares after receiving a complete response letter from the FDA requesting additional clinical data, as well as additional CMC information related to a proposed new dosage form Regeneron was evaluating for gout, which was different than the dosage form approved in the CAPS indication and now being used for pericarditis.
- *Other Indications:* Regeneron conducted a total of 13 clinical trials of rilonacept for the treatment of rheumatoid arthritis, or RA, polymyalgia rheumatica, osteoarthritis, coronary artery disease, systemic juvenile idiopathic arthritis and end-stage renal disease.

In the 21 clinical studies conducted by Regeneron with rilonacept to date, the most common adverse events reported were injection site reactions and upper respiratory tract infections. Across these studies, there

were a total of five serious adverse events, or SAEs, that were assessed by investigators as drug related. Among patients treated with rilonacept there were three SAEs, colitis, gastrointestinal hemorrhage, and drug eruption. One patient treated with placebo experienced cellulitis and another placebo-treated patient died. The largest clinical programs conducted by Regeneron with rilonacept were its Phase 2 and Phase 3 programs for gout flare prevention, which treated a total of 1,886 patients. The most common adverse events reported for the 160 mg dose, the dosage used for the treatment of CAPS, were injection site reactions (15.5% for rilonacept versus 2.6% for placebo) and upper respiratory tract infections (10.3% for rilonacept versus 10.1% for placebo).

Mavrilimumab

Overview

Mavrilimumab is a fully-human monoclonal antibody that antagonizes GM-CSF signaling by binding to the alpha subunit of the GM-CSF receptor. Our lead indication for mavrilimumab is GCA, an inflammatory disease of blood vessels. We have commenced dosing in multiple countries in a double-blind, randomized, placebo-controlled, global Phase 2 proof-of-concept trial for the study of mavrilimumab in GCA. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data in the second half of 2020.

Before we licensed mavrilimumab in 2017, MedImmune, Limited, or MedImmune, was developing mavrilimumab for the treatment of RA.

Mechanism of action

Mavrilimumab is designed to inhibit the signaling of GM-CSF, a growth factor that stimulates the production of certain types of white blood cells. Studies have demonstrated that with GM-CSF overexpression, pathological changes almost always follow. Reported data suggest GM-CSF is a key player in autoinflammation and autoimmunity, as follows:

- GM-CSF enhanced trafficking of myeloid cells through activated endothelium of blood vessels and contributed to monocyte and macrophage accumulation in blood vessels during inflammation;
- GM-CSF promoted activation, differentiation, survival and proliferation of monocytes and macrophages, as well as resident tissue macrophages in inflamed tissues;
- GM-CSF production led to activation of the vasculature and bone marrow and also promoted the differentiation of effector T cells at inflamed sites and draining lymph nodes; and
- GM-CSF regulated the phenotype of antigen-presenting cells in inflamed tissues by promoting the differentiation of infiltrating monocytes into M1 macrophages and monocyte-derived dendritic cells, or MoDCs.

Additionally, GM-CSF has been shown to be a confirmed mediator in RA based on the results from the Phase 2b clinical trial in RA conducted by MedImmune. In this trial, mavrilimumab achieved the co-primary endpoints of change from baseline in disease activity score, or DAS, at week 12 and a response of 20% or greater improvement in the American College of Rheumatology criteria, at week 24. Patients with mavrilimumab showed a statistically significant reduction in DAS scores at all dosages compared to placebo, and significantly more mavrilimumab-treated patients achieved ACR20 at all dosages compared to placebo.

Background and market opportunity for giant cell arteritis

GCA is an inflammatory disease of the blood vessels that strikes older adults and causes headaches, jaw and other muscle claudication, and possible ischemic visual loss. Many of the symptoms and signs of GCA result from involvement of the cranial branches of arteries that originate from the aortic arch, but the disease is systemic, and vascular involvement can be widespread. GCA is characterized by infiltration of monocytes, macrophages and the formation of giant cells (i.e., multinucleated fusions of macrophages). GCA generally occurs in adults over 50 years old with a 3:1 imbalance of women to men. We estimate there to be approximately 75,000 to 150,000 prevalent patients with GCA in the United States with similar prevalence rates for other major markets and believe that the incidence of GCA will increase over time as the population ages.

Current treatment landscape for giant cell arteritis

Glucocorticoids, a type of corticosteroid, are the mainstay for the treatment of GCA because they normalize inflammatory markers and resolve patient symptoms. Many patients receive long courses of this therapy to prevent disease flare-up, which are associated with significant and serious side effects, including glaucoma, fluid retention, hypertension, mood changes, memory changes, other psychological effects, weight gain and diabetes. Up to 80% of patients suffer from glucocorticoid toxicity as a result of GCA treatment.

Despite being effective for some patients, many are unable to wean off of corticosteroids because they continue to experience disease flares as the dose is reduced. In one study cohort published in the literature that followed 106 patients with GCA for 4.5 to 10.1 years, 68 patients (64%) experienced at least one relapse during or after weaning, and 38 patients (36%) experienced two or more. Experimental evidence in mice suggests that corticosteroid treatment does not adequately suppress tissue-infiltrating macrophage function, a key cell type generated and maintained by GM-CSF signaling, and may explain why many patients require long-term chronic treatment and are unable to wean off corticosteroids. We believe by blocking GM-CSF signaling, mavrilimumab may provide additional benefit to these patients by reducing long-term sequelae that results from chronic vessel inflammation.

In addition, tocilizumab, an inhibitor of interleukin-6, or IL-6, is approved in the United States in GCA for use on top of a concomitant corticosteroid taper. However, nearly half of the patients studied in the Phase 3 clinical trial for tocilizumab experienced disease flares during the 52 weeks treatment period that included a 26-week corticosteroid taper. We believe this indicates a persistent unmet medical need.

Our solution

We chose GCA as our first indication for mavrilimumab due to the mechanistic rationale of inhibiting GM-CSF. GM-CSF is a key growth factor for many of these key inflammatory cell types and is found in high concentrations at the site of damage in the vessel wall. We believe these data provide a solid rationale for antagonizing this signaling with mavrilimumab.

Phase 2 clinical trial for GCA

We have commenced dosing in a double-blind, randomized, placebo-controlled, global Phase 2 proof-of-concept trial in multiple countries. In the United States, the FDA initially placed our IND on clinical hold due a request for additional information regarding the 510(k)-cleared delivery device to be used in our Phase 2 clinical trial. The device-related information request did not pertain to pre-clinical toxicology data nor the design of our trial. We have since provided the FDA with the requested information and the IND is now active. We plan for U.S. subjects to be included in the ongoing, global Phase 2 clinical trial, in which

dosing has already commenced in multiple countries. We expect to complete enrollment in this trial in the second half of 2019 and report top-line data in the second half of 2020.

The Phase 2 clinical trial of mavrilimumab for the treatment of GCA is expected to enroll approximately 60 subjects with new-onset and refractory disease. Subjects will be randomized 3:2 to mavrilimumab 150 mg or placebo injected SC once every two weeks co-administered with a corticosteroid taper. Treatment duration is 26 weeks, and the primary efficacy endpoint is time to first flare.

We anticipate that to help inform the risk/benefit profile for the use of mavrilimumab in GCA, we will need to evaluate the effectiveness of different doses as well as mavrilimumab's pharmacokinetic profile. We also intend to initiate research and development activities of mavrilimumab's potential across other various disease states where cells of myeloid phenotype have been implicated by the literature, such as other vasculitides and cardiomyopathies, diseases characterized by barrier dysfunction, other arthropathies or oncologic indications.

Clinical history in rheumatoid arthritis

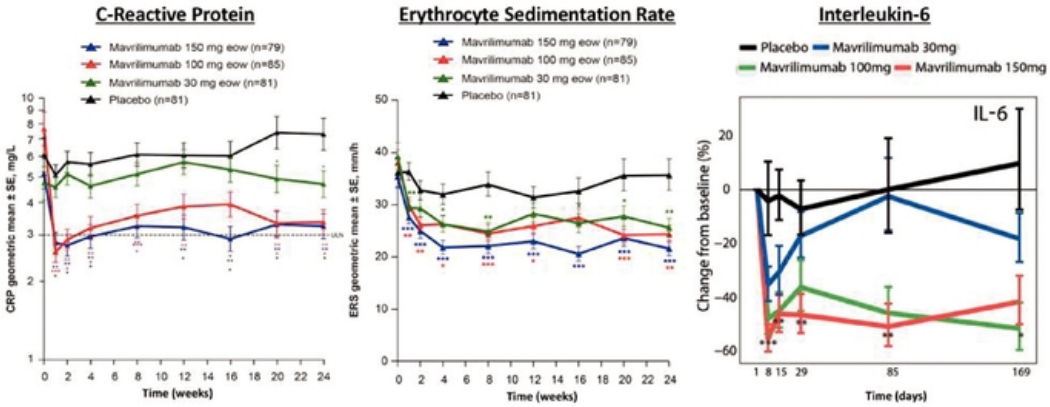
MedImmune had received authorization to conduct clinical trials for rheumatoid arthritis, or RA, in Europe and executed an extensive Phase 1 and Phase 2 clinical program where the company studied mavrilimumab in over 550 patients with RA through Phase 2b. All of MedImmune's European clinical trials achieved their prospectively defined primary endpoints of safety or efficacy.

MedImmune's IND for the clinical development of mavrilimumab for the treatment of RA was initially put on clinical hold in 2010 before human data had been generated due to certain effects that were observed in non-clinical studies, which coincides with a theoretical risk of developing PAP, possibly in the setting of GM-CSF inhibition. Since then, in 2014, the FDA acknowledged that clinical studies in refractory RA may be appropriate based on MedImmune's clinical studies in Europe in which it dosed over 550 RA patients with mavrilimumab with no evidence of PAP attributable to mavrilimumab following long-term administration. MedImmune did not engage in further dialogue with the FDA and withdrew the IND for mavrilimumab for the treatment of RA.

We believe that the trials conducted by MedImmune provide substantial support for the potential of mavrilimumab in autoimmune diseases. In these trials, mavrilimumab was observed to be well-tolerated. The most common adverse event was infection, with all dose groups (30 mg, 100 mg, 150 mg) in a Phase 2b clinical trial reporting similar rates of infection compared to the placebo group. We believe that these safety results provide an accurate early representation of the safety profile of mavrilimumab, which we believe to be at least competitive with and potentially better than existing systemically administered agents for autoimmune diseases.

Mavrilimumab's results from Phase 2b clinical trials in RA have provided important information about its safety and efficacy profile and helped solidify our choice for focusing our development efforts in GCA as a lead indication. In addition to the reductions to the primary endpoint demonstrated in the Phase 2b trials, other markers of inflammation, such as CRP, erythrocyte sedimentation rate, or ESR, and IL-6, were similarly reduced, as shown in the graphs below. CRP, ESR and IL-6 are key markers of disease activity for

GCA. We believe that these results may also provide evidence for mavrilimumab's utility across a broad range of indications with a similar biomarker profile.



KPL-716 overview

KPL-716 is a fully-human monoclonal antibody that targets OSMRb, which mediates signaling of IL-31 and OSM, two key cytokines implicated in inflammation, pruritus and fibrosis. We believe KPL-716 to be the only monoclonal antibody in development that targets both pathways simultaneously. We are initially evaluating KPL-716 for the treatment of a variety of pruritic diseases, including prurigo nodularis and atopic dermatitis, both diseases where OSMRb signaling has been implicated. In September 2018, we announced interim results from the randomized, double-blind, placebo-controlled, single-ascending-dose, sequential-group portion of the Phase 1a/1b clinical trial of KPL-716 in healthy volunteers and in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. In this trial, single IV and SC doses were well-tolerated. Results also provided an early signal of efficacy in reducing pruritus (assessed by the Worst-Itch Numeric Rating Scale), as well as reducing inflammation and disease severity (assessed by EASI), indicative of target engagement. In addition, we observed a reduction of EASI scores in atopic dermatitis subjects after a single dose of KPL-716 in a placebo-controlled exploratory efficacy assessment in subjects who participated in the first portion of the Phase 1a/1b clinical study (20 subjects were randomized 1:1 and received either 7.5 mg/kg of KPL-716 or placebo). We believe these single-dose results provide proof-of-principle for KPL-716's potential to treat a spectrum of IL-31-driven pruritic diseases, and support our plans to advance KPL-716 into multiple chronic pruritic diseases. We plan to initiate an adaptive design Phase 2a/2b clinical trial in prurigo nodularis in the first half of 2019 and expect to report top-line data from the first part of this trial in the first half of 2020. We also plan to initiate an exploratory, pilot Phase 2 clinical trial in the first half of 2019 designed to explore the role of IL-31 and OSM in a number of diseases characterized by chronic pruritus. To help us to understand whether KPL-716 could be a competitive therapeutic in atopic dermatitis, if approved, we are enrolling a repeated-single-dose cohort as an additional part of the Phase 1b clinical trial in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. We expect to report top-line data from this cohort in the second half of 2019.

We acquired the assets relating to KPL-716 from Biogen MA, Inc., or Biogen, in 2016.

Mechanism of action

The OSMRb subunit is an IL-6 type receptor which combines with one of two other subunits to form two distinct cytokine receptors used for the signaling of two different cytokines: IL-31, and OSM. IL-31 produced in the setting of an inflammatory response binds to the IL-31 receptor on keratinocytes, epidermal cells, leading to a sensation of pruritus and further inflammatory responses in the skin. In addition to interacting with IL-31 receptors on keratinocytes, IL-31 also stimulates pruritus directly through IL-31 receptors expressed on unmyelinated C-fibers in the skin responsible for the sensation and transmission of pruritic signaling.

OSM is produced primarily under inflammatory conditions and stimulates dermal fibroblast proliferation and migration as well as synthesis of collagen and glycosaminoglycan in the skin, leading to fibrosis. In addition to these functions, OSM signaling through the type II OSM receptor upregulates interleukin-4, or IL-4, interleukin-13 receptor, or IL-13Ra1, and interleukin-4 receptor, or IL-4Ra, in human skin equivalent cultures, upregulates IL-4Ra in primary human keratinocytes and also impairs expression of filaggrin, loricrin and involucrin (classical "differentiation" markers of the epidermal differentiation complex cluster) in human skin equivalent cultures. These data implicate OSM signaling as important in many autoimmune diseases characterized by barrier dysfunction, fibrosis and inflammation.

KPL-716 inhibits both IL-31 and OSM activities at their respective receptors, potentially disrupting the pruritus, inflammation and fibrosis mediated by these cytokine pathways.

Background and market opportunity for prurigo nodularis, atopic dermatitis and other chronic pruritic diseases

Prurigo nodularis

Prurigo nodularis is a chronic inflammatory skin condition that affects primarily older adults and is characterized by multiple firm and extremely pruritic nodules typically located on the arms and legs. The etiology of prurigo nodularis is largely unknown, however, human biopsy studies have shown that the cytokines IL-31and OSM and the receptor chains IL-31Ra and OSMRb are highly expressed in prurigo nodularis lesions. The pruritus is severe and distressing and can be sudden, sporadic or continuous, worsening with heat, sweating or irritation from clothing. The itching sensation in prurigo nodularis is extreme and often leads to scratching to the point of bleeding, infection or pain. Our market research to-date with physicians and patients highlights the severe and debilitating nature of this disease and the significant levels of unmet need. Multiple physicians have reported suicidal tendencies among their prurigo nodularis patients due to an overwhelming inability to control the unrelenting itch. The exact prevalence of prurigo nodularis is unknown, however, we estimate there to be approximately 300,000 prevalent cases in the United States.

Atopic dermatitis

Atopic dermatitis is a chronic inflammatory skin disease that affects approximately 18 million adults in the United States. Human biopsy studies have shown that the cytokines IL-31 and OSM and the receptor chains IL-31Ra and OSMRb OSMRb are highly expressed in atopic dermatitis lesions. Based upon public data analyses and discussions with physicians and key opinion leaders in the field, we estimate that approximately 300,000 atopic dermatitis patients in the United States are diagnosed with a moderate-to-severe form of this disease that significantly impairs their professional and social life on a daily basis.

Other chronic pruritic diseases

In the exploratory, pilot Phase 2 clinical that we plan to initiate in the first half of 2019, we currently expect to evaluate study populations with chronic idiopathic urticaria, chronic idiopathic pruritus, plaque psoriasis, lichen planus, and lichen simplex chronicus.

- *Chronic idiopathic urticaria.* Chronic idiopathic urticaria is the chronic occurrence of hives without a known cause. We estimate that there are approximately two to three million patients in the United States with chronic idiopathic urticaria. Based on company survey data of over 100 treating physicians, or company survey data, approximately one out of three patients experience pruritus that is refractory to conventional therapies, and among patients treating their chronic idiopathic urticaria with Xolair (omalizumab), 15% to 20% continue to experience pruritus.

- *Chronic idiopathic pruritus.* Chronic idiopathic pruritus is chronic itching without a known cause. Based on company survey data, treating physicians report that there is approximately one patient with idiopathic pruritus for every three atopic dermatitis patients, and approximately 50% of these patients experience symptoms lasting for more than one year and one in three treated patients experience refractory pruritus.

- *Plaque psoriasis.* Plaque psoriasis is the most common form of psoriasis and causes skin lesions with silvery scales. We estimate that there are approximately 12 million patients with plaque psoriasis in the United States with approximately two to three million patients experiencing moderate-to-severe pruritus.

- *Lichen planus.* Lichen planus is a chronic inflammatory and immune-mediated disease that affects the skin, nails, hair, and mucous membranes. We estimate that there are approximately 500,000 patients in the United States with lichen planus. Based on company survey data, treating physicians report that among treated patients with this disease, approximately one in every three experience refractory pruritus.

- *Lichen simplex chronicus.* Lichen simplex chronicus results from chronic itching and scratching, which causes lichenified skin. Based on company survey data, treating physicians report approximately one lichen simplex chronicus patient for every prurigo nodularis patient, which equates to approximately 300,000 addressable patients in the United States, and among treated patients with lichen simplex chronicus, approximately 40% experience refractory pruritus.

Current treatment landscape for prurigo nodularis and atopic dermatitis

Prurigo nodularis

We are not aware of any current FDA-approved therapies for treating prurigo nodularis, and the treatment approach ranges from topical corticosteroids and occlusive steroid containing bandages for more mild patients to systemic corticosteroid, ultraviolet phototherapy and systemic therapies such as thalidomide, methotrexate and cyclosporine for those patients who fail initial treatments. Patients have reported using opioid pain medications to attempt to control the disease in its most severe form.

Atopic dermatitis

Current therapies for atopic dermatitis are generally focused on the topical use of non-biologic small molecules, however, dupilumab (subcutaneously injected antibody directed to inhibiting signaling through IL-4Rα) has recently been approved by the FDA for the treatment of atopic dermatitis.

Our solution

KPL-716 is a fully-human monoclonal antibody that targets two key pathways for the development of pruritus, inflammation and fibrosis through inhibition of OSMRb. Chronic pruritic diseases are often characterized by a complex interplay among pruritus, inflammation and fibrosis. The pathogenesis of chronic pruritic diseases involves interlocking positive feedback loops in which pruritus causes scratch, and scratch causes reactive inflammation through mechanical disruption of the skin architecture. The decline in skin barrier function and resulting bacterial colonization or infection ultimately increase extracellular matrix formation and collagen deposition, leading to fibrosis. Fibrosis then begets more pruritus through disruption and dysregulation of sensory nerve fiber expression.

Current therapies target only one or two aspects of this complex pathophysiology and are inevitably limited in their effectiveness. Targeting only one pathway may address a single aspect of the symptomatology, e.g., pruritus, but not the full spectrum of the pathophysiologic components of the disease. This point is particularly relevant since OSM is upregulated in many chronic inflammatory skin diseases and synergistically interacts with pruritic and inflammatory pathways. Of particular relevance is the central role of OSM in inflammation and barrier function and its autocrine effects on type II OSM receptor in IL-31-dependent epidermal proliferation and remodeling as well as inflammation.

There is a relatively large body of literature linking inflammatory pruritic and inflammatory diseases to both IL-31 and OSM via signaling though OSMRb. KPL-716 has been specifically designed to target both pathways simultaneously and thus KPL-716 may disrupt this pathologic cycle in patients afflicted by prurigo nodularis and atopic dermatitis.

Pre-clinical development

In our pre-clinical development program we have observed favorable pharmacokinetics and toxicology characteristics to support clinical development of KPL-716. KPL-716 has shown signs of efficacy in two non-human primate models. In the first, KPL-716 abrogated the pharmacodynamic marker of pruritus in an IL-31 challenge model. A single three milligram per kilogram dose of KPL-716 substantially reduced scratch counts despite multiple repeated injections of IL-31 over several weeks at concentrations we believe to be supraphysiologic in a disease context. In the second non-human primate model, KPL-716 again abrogated the painful response to an injection to an inflammatory agent called carrageenan through the time period measured after a single infusion of KPL-716, implicating OSM in the inflammatory response. We have conducted pre-clinical toxicology studies for KPL-716 with a no adverse event level of 500 milligrams per kilogram with intravenous dosing.

Phase 1a/1b clinical trial

In early 2017, we filed an IND application and began clinical development with KPL-716 in a Phase 1a/1b clinical trial in healthy volunteers and in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus, respectively.

The first portion of the Phase 1a/1b clinical trial utilized a double-blind, randomized, placebo-controlled, single-ascending-dose, sequential-group design to evaluate the safety, tolerability, pharmacokinetics and immunogenicity of KPL-716 in healthy volunteers and subjects with atopic dermatitis following IV or SC administration. We used the pruritis in atopic dermatitis as a proxy for IL-31-driven pruritic diseases, including prurigo nodularis.

In total, 50 healthy volunteers and 32 subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus received a single dose of KPL-716 or placebo in the Phase 1a/1b clinical trial,

with the top dose of 20 mg/kg IV in healthy volunteers and 7.5 mg/kg IV in subjects with atopic dermatitis. There was a seven-day wash out period of prior therapies for all subjects with atopic dermatitis before treatment, and topical corticosteroids, or TCS, were not allowed through day 28. All subjects were given TSC to use as needed after day 28, and rescue medication was provided for atopic dermatitis flares throughout the study. KPL-716 was well-tolerated, no dose-limiting toxicities were observed, and there were no serious adverse events. All drug-related or potentially drug-related treatment-emergent adverse events, or DR-TEAE, were mild, except for one patient who experienced a moderate DR-TEAE of dizziness.

KPL-716 showed dose-dependent elimination consistent with a target-mediated drug disposition profile and was still detectable at least eight weeks after the high dose of 7.5 mg/kg IV in subjects with atopic dermatitis. We believe the available pharmacokinetic and bioavailability data are supportive of testing once every other week or once monthly SC dosing regimens in subsequent studies of KPL-716.

An exploratory analysis of data in 10 subjects with moderate-to-severe atopic dermatitis receiving a single dose of KPL-716 7.5 mg/kg IV versus ten pooled placebo IV recipients provided an early signal of efficacy for KPL-716 in reducing pruritus, indicative of target engagement. Among these groups, we observed:

- Mean percentage change in weekly-average Worst-Itch Numeric Rating Scale, or WI-NRS, decreased by 40.4% in KPL-716 recipients compared to a 17.6% decrease in placebo recipients at day 28 in the absence of concomitant TCS.
- Mean percentage change in pruritus Visual Analog Scale, or VAS, decreased by 55.4% in KPL-716 recipients compared to a 10.4% decrease in placebo recipients at day 28 in the absence of concomitant TCS.
- 50% of KPL-716 recipients showed a $\geq$ 4-point reduction in weekly-average WI-NRS, compared to 10% of placebo recipients at day 28 in the absence of concomitant TCS.
- The maximum decrease in WI-NRS at day 28 in the absence of concomitant TCS was $\geq$ 8 points (1 subject) in KPL-716 recipients compared to a maximum decrease of 4 points in placebo recipients.
- KPL-716 appeared to show a persistent effect on weekly-average WI-NRS in the period after day 28 through day 56, during which concomitant TCS use was permitted.
- KPL-716 recipients reported a 59.5% decrease in sleep-loss VAS compared to a 2.3% decrease in placebo recipients at day 28 in the absence of concomitant TCS.
- The mean percentage change in EASI (a standardized measure of atopic dermatitis disease severity) decreased by 42.3% in KPL-716 recipients compared to a 25% decrease in placebo recipients at Day 28 in absence of concomitant TCS.

We continue to enroll a repeated-single-dose cohort as an additional part of the Phase 1b clinical trial in the United States and Canada. In this double-blind, randomized, placebo-controlled study, subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus receive 360 mg or placebo via SC injection of KPL-716 once weekly for 12 weeks. The clinical study is designed to provide similar and longer-term exposures in order to evaluate safety and chronic efficacy data on both pruritus and inflammation and disease severity (assessed by EASI) response markers compared to the single-dose IV cohort in the Phase 1b clinical trial. We expect to report top-line data from this cohort in the second half of 2019.

Adaptive Design Phase 2a/2b Clinical Trial in Prurigo Nodularis

Based on the results from the single-dose IV Phase 1b results in atopic dermatitis subjects, we intend to initiate an adaptive design Phase 2a/2b randomized, double-blind, placebo-controlled clinical trial to investigate the efficacy, safety, tolerability, PK and immunogenicity of KPL-716 administered SC in subjects with moderate to severe prurigo nodularis experiencing moderate to severe pruritus. We expect that the Phase 2a portion of the study will enroll up to approximately 100 subjects with moderate to severe prurigo nodularis experiencing moderate to severe pruritus and will include two arms: one active arm and one placebo arm. A total of 16 doses of study drug, administered subcutaneously, once-weekly for 16 weeks, are planned during the treatment period to assess potential proof of concept in prurigo nodularis on the endpoints of pruritus, sleep, quality of life and disease severity, with dosing to achieve maximum or near-maximum steady-state exposures. The primary and key secondary endpoints, which focus on pruritus, are set at 8 weeks. Other secondary endpoints will explore the impact of KPL-716 versus placebo on pruritus, sleep, quality of life and disease severity over time (at each week of the study treatment period up to and including week 16).

We may perform an interim analysis of the Phase 2a portion of the study to assess for an early signal of efficacy. Depending on the results of this interim analysis, enrollment into the Phase 2b portion of the study could be initiated prior to completion of the Phase 2a portion of the study. Subjects enrolled in the Phase 2a portion would remain in the Phase 2a portion.

The Phase 2b portion of the study, if enrolled, will include up to approximately 300 subjects with moderate to severe prurigo nodularis experiencing moderate to severe pruritus and will have five arms: four active arms, testing different dose levels or dosing regimens of KPL-716, and one placebo arm. The primary endpoint of the Phase 2b portion will also focus on pruritus at week 8, with other secondary endpoints evaluating pruritus, sleep, quality of life and disease severity over time.

Exploratory Multi-Indication, Pilot Phase 2 Clinical Trial

We believe there are multiple chronic pruritic diseases where IL-31 and OSM may play a role in disease pathology. In the first half of 2019, we plan to initiate an exploratory, multi-indication, randomized, double-blind, placebo-controlled, pilot clinical trial designed to (1) explore the role of IL-31 and OSM in a number of diseases characterized by chronic pruritus seen by dermatologists or allergists and (2) investigate the efficacy, safety and tolerability of KPL-716 in reducing the moderate to severe pruritus experienced by these subjects. We currently expect the study populations to be chronic idiopathic urticaria, chronic idiopathic pruritus, plaque psoriasis, lichen planus, and lichen simplex chronicus.

The trial will be comprised of multiple cohorts, each for a different study population. Each cohort will be an independent sub-study assessing each study population for presence of an IL-31 or OSM protein or ribonucleic acid signature via biopsy and investigating the efficacy, safety and tolerability of KPL-716 administered SC in reducing pruritus in these populations. Investigators and subjects will remain blinded throughout the study. We expect each cohort to enroll up to approximately 26 subjects with each subject experiencing WI-NRS of seven or above at screening. A loading dose of KPL-716 (720 mg) or matching placebo will be administered on day 1, followed by single, weekly SC injections of KPL-716 (360 mg) or matching placebo for the next seven weeks, with the goal of achieving maximum or near maximum exposures at steady state. The goal of this exploratory study is to identify chronic pruritic conditions where IL-31 and/or OSM may be playing a role and assess the presence or absence of reduction in pruritus after KPL-716 treatment.

Pre-clinical development

KPL-045

KPL-045 is a fully-human monoclonal antibody that is designed to inhibit the CD30-CD30 ligand interaction, a co-stimulatory signal helpful in activating and sustaining memory T-cells. The majority of the therapeutics in development modulating the CD30-CD30 ligand interaction are depleting or conjugated to a toxin for the use in hematological malignancies. To our knowledge, KPL-045 is the only non-depleting antibody targeting primarily autoimmune disease in active clinical development. In August 2017, we licensed this antibody from Novo Nordisk.

In pre-clinical development, of KPL-045 has been observed to have a favorable pharmacokinetic profile to support further development. KPL-045 has showed single-digit nanomolar potency against both human and cynomolgus non-human primate CD30L. We are continuing our pre-clinical activities in KPL-045 in inflammatory diseases driven by T-cell-dependent autoantibody generation and dysregulated T_H effector memory responses and expect to file an IND with the FDA for this program in the second half of 2019 and to initiate a Phase 1 clinical trial in the first half of 2020.

KPL-404

KPL-404 is a humanized monoclonal antibody that is designed to inhibit the CD40-CD40 ligand interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching. We have a license to conduct research and development on KPL-404 from Primatope Therapeutics, Inc., or Primatope, the company that owns or controls the intellectual property related to KPL-404. In January 2019, we exercised our exclusive option to acquire all outstanding capital stock of Primatope, which, subject to a supplemental due diligence period, we would expect to close within 60 days of the exercise of our option. If we close the transaction, we will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10.0 million as well as potential milestone payments of up to $8.0 million ($3.0 million of which has been achieved as of the date of this prospectus and would be payable at closing) payable in a combination of cash and our Class A common shares. If we do not close the transaction, our license to intellectual property controlled by Primatope to research, develop and manufacture KPL-404 will terminate, in which case we would cease the development of KPL-404.

In pre-clinical development, KPL-404 has been observed to have a favorable pharmacokinetic and toxicology profile to support further development. KPL-404 has been effective in multiple non-human primate models of organ transplant rejection, as well as in multiple T-cell dependent antibody response models. We are continuing our pre-clinical activities in KPL-404 in T-cell dependent, B-cell mediated diseases, and expect to file an IND application with the FDA for this program in the second half of 2019 and to initiate a Phase 1 clinical trial in the first half of 2020.

Discovery activities

We have initiated internal discovery activities directed toward wholly owned molecules for the treatment of autoinflammatory and autoimmune disease targets where we believe there to be a strong mechanistic rationale and clear differentiation from existing approved agents or those in development.

License and acquisition agreements

License agreement with Regeneron

In September 2017, we entered into a license agreement with Regeneron, or the Regeneron Agreement. Pursuant to the Regeneron Agreement, Regeneron granted us an exclusive license under certain intellectual property rights controlled by Regeneron to develop and commercialize rilonacept worldwide, aside from Israel, Egypt, Turkey and select countries in the Middle East and northern Africa, which we refer to collectively as the Excluded Territory. In the United States and Japan, our license is initially for all indications other than those involving local administration to the eye or ear, oncology, deficiency of the interleukin-1 receptor antagonist, or DIRA, and CAPS. If we are successful in receiving marketing approval for rilonacept in the United States for a new indication, the scope of the license granted to us will automatically expand to include DIRA and CAPS in the United States and Japan, and we will assume the sales and distribution of rilonacept in these additional indications. Outside the Unites States and Japan, our license is for all indications other than local application to the eye or ear, oncology, CAPS, DIRA and certain periodic fever syndromes set forth in the Regeneron Agreement, collectively the Excluded Indications. Under the Regeneron Agreement, we are obligated to use commercially reasonable efforts to develop and commercialize rilonacept outside of the Excluded Indications in our territory. Upon receiving positive data in a Phase 3 clinical trial, Regeneron will transfer the BLA for rilonacept to us.

We made an upfront payment of $5.0 million to Regeneron and are obligated to make regulatory milestone payments of up to $27.5 million in the aggregate. Thereafter, we have agreed to evenly split profits on our sales of rilonacept with Regeneron after deducting certain commercialization expenses subject to specified limits.

Regeneron has a right of first negotiation over our engagement of third parties to support our promotional activities in excess of a specified level and over the assignment or sale of our rights to any product we develop under the Regeneron Agreement to a third-party. Furthermore under certain circumstances, we will need Regeneron's prior consent to assign our rights under the Regeneron Agreement.

The Regeneron Agreement will expire on the date on which we, our affiliates or sublicensees are no longer developing or commercializing any product containing rilonacept. We may terminate the agreement for convenience at any time after the date that is 18 months after the effective date of the agreement with 180 days' written notice or one year's written notice if we terminate the agreement following U.S. marketing approval of a rilonacept product developed by us. We may also terminate with three months' written notice if we reasonably determine that rilonacept is unsafe in the indications we are pursuing. Regeneron may terminate the agreement if there is a consecutive twelve (12) month period during which we do not conduct any material development or commercialization activities or we do not grant a sublicense to a third-party to do so, or if we challenge Regeneron's patent rights in any country in our territory. Either party may terminate the agreement in the event of a material breach by the other party that remains uncured for 90 days (or 30 days for payment-related breaches), or by either party due to the insolvency or bankruptcy of the other party.

We have also entered into a clinical supply agreement with Regeneron, or the Supply Agreement. Pursuant to the Supply Agreement, Regeneron has the exclusive right to manufacture and supply all of our requirements of rilonacept for clinical development. If Regeneron determines to discontinue the supply of rilonacept to us, it must use its reasonable efforts to transfer all relevant documentation, materials and technology necessary for the manufacture of rilonacept to us or our designee. The Supply Agreement terminates upon the termination of the Regeneron Agreement or the transfer of technology related to the bulk manufacture of rilonacept.

License agreement with MedImmune

In December 2017, we entered into a license agreement with MedImmune, or the MedImmune Agreement. Pursuant to the MedImmune Agreement, MedImmune granted us an exclusive, worldwide license under certain intellectual property rights controlled by MedImmune to make, use, develop and commercialize mavrilimumab and any other product containing an antibody to the GM-CSF receptor alpha that is covered by certain MedImmune patent rights for all indications. We also acquired non-exclusive licenses to other MedImmune technology for use in exploiting licensed products. We may sublicense these rights subject to consent of MedImmune and any applicable licensors of rights under which we are licensed. We also acquired reference rights to relevant manufacturing and regulatory documents, and existing inventory of mavrilimumab drug substance. We must use commercially reasonable efforts to develop and commercialize the licensed products.

We made an upfront payment of $8.0 million to MedImmune and are obligated to make future clinical, regulatory and initial sales milestone payments of up to $72.5 million in the aggregate for the first two indications, including a milestone payment of $10.0 million which we paid upon the occurrence of a specified regulatory milestone, and clinical and regulatory milestone payments of up to $15.0 million in the aggregate for each subsequent indication. We are also obligated to make milestone payments to MedImmune of up to $85.0 million upon the achievement of annual net sales thresholds up to, but excluding, $1.0 billion in annual net sales as well as additional milestone payments aggregating up to $1.1 billion upon the achievement of additional annual net sales thresholds starting at $1.0 billion and higher. Commencing on the first commercial sale of licensed products, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. We must pay such royalties on a product-by-product and country-by-country basis until the latest to occur of the expiration of licensed patents, the expiration of regulatory exclusivity or the tenth anniversary of first commercial sale of such product in such country.

In countries where licensed patents have issued, the statutory expiration date is 2027, not including any patent term extensions or adjustments. While the current expected patent expiration dates are known in countries where licensed patents have issued, these expiration dates are subject to significant uncertainty. For example, the patents may be challenged, and accordingly, the relevant expiration dates could be shortened. In addition, as we continue to file and prosecute patent applications related to mavrilimumab, the granting of pending applications or future patent applications could extend the relevant statutory expiration dates beyond 2027. The expiration date of regulatory exclusivity is determined on a country-by country-basis if the applicable product is approved in such country and if any applicable regulatory exclusivity applies and is granted. The actual expiration date of any such regulatory exclusivity, however, is subject to significant uncertainty. For instance, the applicable regulatory exclusivity period is often triggered by the date a product candidate obtains regulatory approval, and we cannot predict with any certainty whether and if so, when, the applicable product would receive regulatory approval in any given jurisdiction. Furthermore, the type, scope and duration of such exclusivities will vary on a country-by-country basis depending on the jurisdictions in which a product candidate is approved and the particular regulatory exclusivity for which the product is eligible as of the time of approval. For example, in the United States, a reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, which means that the FDA cannot make effective the approval of a biosimilar product that references the biologic product until 12 years from the date on which the reference product was first licensed. In the European Union, new products authorized for marketing may qualify for eight years of data exclusivity and an additional two years of market exclusivity upon marketing authorization. The two-year period may be extended to three years if during the first eight years a new therapeutic indication with significant clinical benefit over existing therapies is approved. Furthermore, if a product

candidate that has received orphan designation is subsequently approved for the disease or condition for which it has such designation, the product may be entitled to orphan drug exclusivity, which generally grants seven years of market exclusivity in the United States and up to 10 years of market exclusivity in the European Union, and such period may run contemporaneously with the other exclusivities that may apply. In the European Union, an orphan product can also obtain an additional two years of market exclusivity for pediatric studies. In the United States, an additional six-month period of pediatric exclusivity may be available as an extension to any existing non-patent regulatory exclusivity period if the sponsor has conducted and submitted pediatric studies in response to a written request from the FDA. Additionally, our eligibility for regulatory exclusivity may depend in part on the indications for which we seek regulatory approval of our product candidates, which may depend on the data we receive from our clinical studies, and accordingly, may change over time, and the laws and regulations governing regulatory exclusivity may change in various jurisdictions as the political focus on drug exclusivity increases. For risk related to regulatory exclusivity matters, see "Risk factors—Risks related to product development and regulatory approval."

The MedImmune Agreement will remain in effect until the expiration of the royalty term in all countries for all licensed products. The MedImmune Agreement may be terminated earlier at any time by us with at least 90 days' prior notice, by either party in the event of material breach by the other party that remains uncured for 90 days, by either party for insolvency or bankruptcy of the other party, or immediately by MedImmune if we challenge the licensed patents.

Biogen asset purchase agreement

In September 2016, we completed the acquisition of certain assets of Biogen pursuant to an asset purchase agreement, or the Biogen Agreement. Pursuant to the Biogen Agreement, we acquired all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716 and other antibodies covered by certain patent rights, together the Acquired Assets, including patents and other intellectual property rights, clinical data, certain contracts, know-how and inventory. In addition, Biogen granted us a non-exclusive, sublicensable, worldwide license to certain background patent rights related the KPL-716 program. Under the Biogen Agreement, we are obligated to use commercially reasonable efforts to develop and commercialize the Acquired Assets.

Under the Biogen Agreement, we made an upfront payment of $11.5 million and a technology transfer payment of $0.5 million to Biogen. In addition, we made a milestone payment of $4.0 million during the year ended December 31, 2017 associated with the achievement of a specified clinical milestone event. We are also obligated to make future milestone payments for each antibody product that includes the Acquired Assets, or an Antibody Product, of up to $325.0 million in the aggregate upon the achievement of specified milestones. These milestone payments relate to multiple indications for an Antibody Product, and are comprised of up to $175.0 million in the aggregate upon achievement of specified clinical and regulatory milestone events and $150.0 million in the aggregate upon the achievement of specified annual net sales thresholds. Commencing on the first commercial sale of an Antibody Product, we are obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens. We must pay such royalties on a product-by-product and country-by-country basis until the latest to occur of the expiration of patents that cover an Antibody Product, the expiration of regulatory exclusivity or the tenth anniversary of first commercial sale of such product in such country. We have also agreed to pay certain obligations under third-party contracts retained by Biogen that relate to KPL-716.

In countries where patents covering Antibody Products have issued, the statutory expiration date is 2034, not including any patent term extensions or adjustments. While the current expected patent expiration dates are known in countries where licensed patents have issued, these expiration dates are subject to significant uncertainty. For example, the patents may be challenged, and accordingly, the relevant expiration dates could be shortened. In addition, as we continue to file and prosecute patent applications related to Antibody Products, the granting of pending applications or future patent applications could extend the relevant statutory expiration dates beyond 2034. The expiration date of regulatory exclusivity is determined on a country-by country-basis if the applicable product is approved in such country and if any applicable regulatory exclusivity applies and is granted. The actual expiration date of any such regulatory exclusivity, however, is subject to significant uncertainty. For instance, the applicable regulatory exclusivity period is often triggered by the date a product candidate obtains regulatory approval, and we cannot predict with any certainty whether and if so, when, the applicable product would receive regulatory approval in any given jurisdiction. Furthermore, the type, scope and duration of such exclusivities will vary on a country-by-country basis depending on the jurisdictions in which a product candidate is approved and the particular regulatory exclusivity for which the product is eligible as of the time of approval. For example, in the United States, a reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, which means that the FDA cannot make effective the approval of a biosimilar product that references the biologic product until 12 years from the date on which the reference product was first licensed. In the European Union, new products authorized for marketing may qualify for eight years of data exclusivity and an additional two years of market exclusivity upon marketing authorization. The two-year period may be extended to three years if during the first eight years a new therapeutic indication with significant clinical benefit over existing therapies is approved. Furthermore, if a product candidate that has received orphan designation is subsequently approved for the disease or condition for which it has such designation, the product may be entitled to orphan drug exclusivity, which generally grants seven years of market exclusivity in the United States and up to 10 years of market exclusivity in the European Union, and such period may run contemporaneously with the other exclusivities that may apply. In the European Union, an orphan product can also obtain an additional two years of market exclusivity for pediatric studies. In the United States, an additional six-month period of pediatric exclusivity may be available as an extension to any existing non-patent regulatory exclusivity period if the sponsor has conducted and submitted pediatric studies in response to a written request from the FDA. Additionally, our eligibility for regulatory exclusivity may depend in part on the indications for which we seek regulatory approval of our product candidates, which may depend on the data we receive from our clinical studies, and accordingly, may change over time, and the laws and regulations governing regulatory exclusivity may change in various jurisdictions as the political focus on drug exclusivity increases. For risk related to regulatory exclusivity matters, see "Risk factors—Risks related to product development and regulatory approval."

Under the Biogen Agreement, Biogen has a time-limited right of first negotiation to purchase the assets we acquired from Biogen or obtain a license to exploit Antibody Products, in each case, in the event we decide to sell the acquired assets, including through the sale of our company, or out-license the rights to the Antibody Products.

The Biogen Agreement will remain in effect until expiration of all payment obligations in all countries related to the last antibody product subject to the Biogen Agreement. The Biogen Agreement may be terminated by us with 90 days' prior notice, by either party in the event of a material breach by the other party that remains uncured for 90 days (or 30 days for payment-related breaches) or by both parties upon mutual consent. In the event of a termination, the Acquired Assets, including certain licenses and rights related thereto, will revert to Biogen, and, upon written request by Biogen, we are required to grant to Biogen an exclusive, worldwide, sub-licensable license to certain of our intellectual property related to the Acquired Assets, including know-how and patent rights.

Manufacturing

We do not currently own or operate any large-scale manufacturing facilities. Although we built small-scale manufacturing facilities to produce drug substance to support certain of our pre-clinical studies and certain of our Phase 1 clinical studies, we rely on third parties to manufacture all of our product candidates. We have entered into a clinical supply agreement with Regeneron to manufacture and supply rilonacept for our clinical trials. Regeneron has also agreed to provide commercial drug material until at least the later of four years after U.S. marketing approval or seven years after the effective date of the agreement.

We believe that we have sufficient quantities of drug substance to supply our planned Phase 2 clinical trial of mavrilimumab for the treatment of GCA. We also acquired a certain amount of finished mavrilimumab drug product that we plan to use in this clinical trial, and we intend to enter into a fill/finish supply agreement with a contract manufacturing organization, or CMO, to produce additional finished mavrilimumab drug product from our current inventory of drug substance. We plan to transfer the manufacturing process of mavrilimumab to, and enter into an agreement with, a CMO to produce mavrilimumab drug substance beyond our existing inventory for other clinical trials, including any Phase 3 clinical trial, and eventual commercialization of mavrilimumab, if approved. There are certain components, for example, media and feed, used to produce our current mavrilimumab inventory that we will not use in our future manufacturing process. We and any CMO that we enter into agreement with to manufacture mavrilimumab will need to find alternative components to replace the media and feed that had been used by MedImmune to date in the manufacture of mavrilimumab.

We acquired a certain amount of KPL-716 drug substance from Biogen from which we produced KPL-716 drug product using a CMO. In addition, we have engaged CMOs to manufacture KPL-716 drug substance and product for further clinical development activities. We intend to continue using CMOs to develop our manufacturing process and scale-up for any future clinical trials and eventual commercialization of KPL-716, if approved.

We are currently using CMOs to produce our pre-clinical product candidates, KPL-045 and KPL-404, for our planned IND-enabling studies, but we intend to produce these product candidates for Phase 1 studies in our own small-scale manufacturing facilities. Longer-term, we expect to use CMOs to produce these product candidates for later-phase clinical studies and eventual commercialization, if approved.

We require all of our CMOs to conduct manufacturing activities in compliance with current good manufacturing practice, or cGMP, requirements. We have assembled a team of experienced employees and consultants to provide the necessary technical, quality and regulatory oversight of our CMOs. We currently rely solely on these third-party manufacturers for scale-up and process development work and to produce sufficient quantities of product candidate for use in pre-clinical studies and clinical trials. Although we have established our own small-scale manufacturing capabilities to support certain pre-clinical and early clinical-stage production of product candidates, we intend to continue to rely on third-party manufacturers for clinical and commercial supply of our product candidates. We anticipate that these CMOs will have the capacity to support both clinical supply and commercial-scale production, but we do not have any formal agreements at this time with any of these CMOs to cover commercial production. We also may elect to pursue additional CMOs for manufacturing supplies of drug substance and finished drug product in the future.

Commercial operations

Our team is experienced in commercial leadership and we intend to expand our capabilities in parallel with the development path of our product candidates. If the FDA approves rilonacept for recurrent pericarditis, we intend to market and commercialize rilonacept in the United States by developing our own sales, marketing and medical affairs organizations targeting a subset of cardiologists and rheumatologists currently treating pericarditis. For our other product candidates, we intend to establish commercialization strategies for each as we approach potential marketing approval and, due to the specialization among physicians treating the indications we are targeting, we expect to be able leverage our then-existing sales, marketing and medical affairs organizations.

Competition

The biotechnology and pharmaceutical industries are characterized by rapidly advancing technologies, intense competition and a strong emphasis on proprietary products. We face potential competition from many different sources, including pharmaceutical and biotechnology companies, academic institutions and governmental agencies and public and private research institutions. Any product candidates that we successfully develop and commercialize, including rilonacept, mavrilimumab and KPL-716, may compete with existing products and new products that may become available in the future.

Our competitors may have significantly greater financial resources, established presence in the market, expertise in research and development, manufacturing, pre-clinical and clinical testing, obtaining regulatory approvals and reimbursement and marketing approved products than we do. These competitors also compete with us in recruiting and retaining qualified scientific, sales, marketing and management personnel, establishing clinical trial sites and patient registration for clinical trials, as well as in acquiring technologies complementary to, or necessary for, our programs. Smaller or early-stage companies may also prove to be significant competitors, particularly through collaborative arrangements with large and established companies.

The key competitive factors affecting the success of rilonacept, mavrilimumab and KPL-716, and any other product candidates that we develop, if approved, are likely to be their efficacy, safety, convenience, price, the level of generic competition and the availability of reimbursement from government and other third-party payors. Our commercial opportunity for any of our product candidates could be reduced or eliminated if our competitors develop and commercialize products that are more effective, have fewer or less severe side effects, are more convenient or are less expensive than any products that we may develop. Our competitors also may obtain FDA or other regulatory approval for their products more rapidly than we may obtain approval for ours. In addition, our ability to compete may be affected in many cases by insurers or other third-party payors seeking to encourage the use of generic products.

We are aware of the following products currently marketed or in clinical development for the treatment of the diseases that we are initially targeting:

Rilonacept

We are not aware of any therapies currently approved by the FDA for the treatment of recurrent pericarditis, our lead indication for rilonacept. Anakina (KINERET), produced by Sobi, Inc., is an FDA-approved agent that inhibits IL-1a and IL-1b signaling and is approved for RA and CAPS. Canakinumab (ILARIS), produced by Novartis Pharmaceuticals Corporation, is a monoclonal antibody which inhibits IL-1b signaling and is approved for use in CAPS, tumor necrosis factor receptor associated period syndrome, hyperimmunoglobulin D syndrome, familiar Mediterranean fever and active systemic juvenile idiopathic

arthritis. There are also other therapies modulating IL-1a and/or IL-1b which are in various stages of clinical development for diseases other than recurrent pericarditis from AbbVie, Inc., or AbbVie, XBiotech Inc. and Handok Inc.

Mavrilimumab

Tocilizumab (ACTEMRA), produced by Hoffmann—La Roche AG, or Roche, and Chugai Pharmaceutical Co., Ltd., is an IL-6 inhibitor that is approved by the FDA for the treatment of GCA on top of a concomitant corticosteroid taper. There are also five other programs in clinical development that modulate GM-CSF signaling from GlaxoSmithKline plc, or GSK, Izana Bioscience Ltd., Morphotek, Inc., Janssen and Humanigen, Inc. In addition, Eli Lilly and AbbVie are conducting clinical trials for oral janus kinase inhibitors. Furthermore, Sanofi S.A. and Regeneron intend to initiate a Phase 3 clinical trial with their anti-IL-6 program in 2018, Novartis International AG, plans to start a trial with secukinumab (Cosentyx) and Janssen's ustekinumab (STELARA) is being trialed in two small studies for GCA.

KPL-716

We are not aware of any therapies currently approved by the FDA for the treatment of prurigo nodularis. Menlo Therapeutics Inc., Vanda Pharmaceuticals Inc., Trevi Therapeutics, Inc. and Galderma SA, or Galderma, have programs in various stages of clinical development for the treatment of prurigo nodularis.

The FDA recently approved Regeneron's dupilumab, an antibody that inhibits signaling through the interleukin 4 receptor, to treat atopic dermatitis. Other companies currently developing systemic therapies for atopic dermatitis include Roche, Dermira, Inc., Galderma, Asana BioSciences, LLC, Eli Lilly and Co., Pfizer Inc., AbbVie, Glenmark Pharmaceuticals Ltd., GSK, LEO Pharma Inc., Incyte Corporation, Dermavant Sciences, Inc., Novartis International AG, MedImmune, and AnaptsysBio, Inc.

Intellectual property

Our success depends in part on our ability to obtain and maintain proprietary protection for our drug candidates, manufacturing and process discoveries, and other know-how, to operate without infringing the proprietary rights of others, and to prevent others from infringing our proprietary rights. We plan to protect our proprietary position using a variety of methods, which include pursuing U.S. and foreign patent applications related to our proprietary technology, inventions and improvements, including compositions of matter, drug product formulations, and methods-of-use, that are important to the development and implementation of our business. For example, we or our licensors have or are pursuing patents covering the composition of matter for each of our product candidates and we generally pursue patent protection covering methods-of-use for each clinical program. We also rely on trade secrets, know-how, continuing technological innovation and potential in-licensing opportunities to develop and maintain our proprietary position.

We have a field-specific exclusive license under the Regeneron Agreement to granted patents and pending applications in the United States and numerous foreign jurisdictions relating to rilonacept. As of December 31, 2018, the patent rights in-licensed under the Regeneron Agreement relating to our program include one granted patent in the United States and 54 patents granted in foreign jurisdictions, including Canada, Australia, Brazil and selected countries in Europe and Asia. In addition, the patent rights in-licensed under the Regeneron Agreement relating to our program include patent applications that are pending in the United States. A U.S. patent covering rilonacept as a composition of matter has a statutory expiration date in 2019, not including patent term adjustment, and relevant foreign counterparts are expected to expire between 2019 and 2023, in each case, not including any patent term extensions. If we

are successful in obtaining regulatory approval of rilonacept for the treatment of recurrent pericarditis and receive orphan designation, we would rely on orphan exclusivity, which generally grants seven years of marketing exclusivity in the United States and 10 years of marketing exclusivity in Europe. See "—License agreement with Regeneron" above for additional information on our rights under the Regeneron Agreement.

We have an exclusive license under the MedImmune Agreement to granted patents and pending patent applications in the United States and numerous foreign jurisdictions relating to mavrilimumab. These patents and patent applications cover mavrilimumab as a composition of matter and its use. As of December 31, 2018, the patent rights in-licensed under the MedImmune Agreement relating to our program include three granted patents in the United States and 106 patents granted in foreign jurisdictions, including Canada, Australia and selected countries in Europe and Asia. In addition, the patent rights in-licensed under the MedImmune Agreement relating to our program include patent applications that are pending in the United States and selected countries in Asia and Latin America. The composition of matter patents for mavrilimumab generally have statutory expiration dates in 2027, although the term of some U.S. patents may be longer due to patent term adjustment to compensate for delays during the patent prosecution process. Patent term extension could extend the expiration date of one patent in the United States and patents in certain other jurisdictions, each in accordance with applicable law. There can be no assurances that patents will issue from any pending patent applications. See "—License agreement with MedImmune" above for additional information on our rights under the MedImmune Agreement.

We own, via our acquisition of certain assets from Biogen, granted patents and pending patent applications in the United States and numerous foreign jurisdictions relating to KPL-716. These patents and patent applications cover KPL-716 as a composition of matter and its use. As of December 31, 2018, the patent rights acquired from Biogen include three patents granted in the United States and 30 patents granted in foreign jurisdictions, including Australia, Mexico and selected countries in Europe and Asia. In addition, the patent rights acquired from Biogen include patent applications pending in the United States, Europe, Canada, and selected countries in Asia. The issued composition of matter patents for KPL-716 have statutory expiration dates in 2034. Patent term extension could extend the expiration date of one patent in the United States and patents in certain other jurisdictions, each in accordance with applicable law. There can be no assurance that patents will issue from any of our pending patent applications. See "—Biogen asset purchase agreement" above for additional information on our rights under the Biogen Agreement.

The term of individual patents depends upon the legal term for patents in the countries in which they are obtained. In most countries, including the United States, the patent term is 20 years from the earliest filing date of a non-provisional patent application. In the United States, a patent's term may be lengthened by patent term adjustment, which compensates a patentee for administrative delays by the USPTO, in examining and granting a patent, or may be shortened if a patent is terminally disclaimed over an earlier expiring patent. In certain countries, the term of a patent that covers a drug product may also be eligible for patent term extension when regulatory approval is granted, provided the legal requirements are met. In the future, if and when our drug candidates receive approval by the FDA or foreign regulatory authorities, we expect to apply for patent term extensions on issued patents covering those drugs, depending upon the length of the clinical trials for each drug and other factors. There can be no assurance that any of our pending patent applications will issue or that we will benefit from any patent term extension or favorable adjustment to the term of any of our patents.

Government regulation

Government authorities in the United States at the federal, state and local level and in other countries and jurisdictions, including the European Union, extensively regulate, among other things, the research, development, testing, manufacture, quality control, approval, labeling, packaging, storage, record-keeping, promotion, advertising, distribution, post-approval monitoring and reporting, marketing and export and import of drug products, such as rilonacept, mavrilimumab and our other product candidates. Generally, before a new drug can be marketed, considerable data demonstrating its quality, safety and efficacy must be obtained, organized into a format specific for each regulatory authority, submitted for review and approved by the regulatory authority.

U.S. government regulation of biological products

In the United States, the FDA regulates biologics under the Federal Food, Drug, and Cosmetic Act, or FDCA, and the Public Health Service Act, or PHSA, and their implementing regulations. Products are also subject to other federal, state and local statutes and regulations. The process of obtaining regulatory approvals and the subsequent compliance with applicable federal, state, local and foreign statutes and regulations requires the expenditure of substantial time and financial resources. Failure to comply with the applicable U.S. requirements at any time during the product development process, approval process or after approval, may subject an applicant to a variety of administrative or judicial sanctions, such as the FDA's refusal to approve pending biologic license applications, or BLAs, withdrawal of an approval, imposition of a clinical hold, issuance of warning letters, product recalls, product seizures, total or partial suspension of production or distribution, injunctions, fines, refusals of government contracts, restitution, disgorgement of profits or civil or criminal penalties.

The process required by the FDA before a biologic may be marketed in the United States generally involves the following:

- Completion of extensive pre-clinical studies and tests in accordance with applicable regulations, including Good Laboratory Practice, or GLP, regulations and applicable requirements for the humane use of laboratory animals or other applicable regulations;
- Submission to FDA of an IND which must become effective before human clinical trials may begin;
- Approval by an independent institutional review board, or IRB, or ethics committee at each clinical trial site before each trial may be initiated;
- Performance of adequate and well-controlled human clinical trials in accordance with applicable IND regulations, good clinical practices, or GCPs, and other clinical-trial related regulations to evaluate the safety and efficacy of the investigational product for each proposed indication;
- Submission to FDA of a BLA for marketing approval that includes substantive evidence of safety, purity, and potency from results of pre-clinical testing and clinical trials;
- A determination by FDA within 60 days of its receipt of a BLA to accept the filing for review;
- Satisfactory completion of one or more FDA pre-approval inspections of the manufacturing facility or facilities where the biologic will be produced to assess compliance with cGMPs to assure that the facilities, methods and controls used in product manufacture are adequate to preserve the biologic's identity, strength, quality and purity;

- Potential FDA audit of the pre-clinical and/or clinical trial sites that generated the data in support of the BLA;
- Payment of user fees for FDA review of the BLA; and
- FDA review and approval of the BLA, including satisfactory completion of an FDA advisory committee review, if applicable, prior to any commercial marketing or sale of the product in the United States.

Pre-clinical studies

Before testing any biological product candidate, including our product candidates, in humans, the product candidate must undergo rigorous pre-clinical testing. The pre-clinical development stage generally involves laboratory evaluations of the chemistry, formulation and stability of the product candidate, as well as trials to evaluate toxicity in animals, which support subsequent clinical testing. The conduct of the pre-clinical tests must comply with federal regulations and requirements, including GLP regulations. The sponsor must submit the results of the pre-clinical studies, together with manufacturing information, analytical data, any available clinical data or literature and a proposed clinical protocol, to the FDA as part of the IND. An IND is a request for authorization from the FDA to administer an investigational product to humans, and must become effective before human clinical trials may begin. Some pre-clinical testing may continue even after the IND is submitted. An IND automatically becomes effective 30 days after receipt by the FDA, unless before that time the FDA raises concerns or questions related to one or more proposed clinical trials and places the clinical trial on a clinical hold. In such a case, the IND sponsor and the FDA must resolve any outstanding concerns before the clinical trial can begin. As a result, submission of an IND may not result in the FDA allowing clinical trials to commence.

Clinical trials

The clinical stage of development involves the administration of the investigational product to healthy volunteers or patients under the supervision of qualified investigators, generally physicians not employed by, or under control of, the trial sponsor, in accordance with GCPs, which include the requirement that all research subjects provide their informed consent for their participation in any clinical trial. Clinical trials are conducted under protocols detailing, among other things, the objectives of the clinical trial, dosing procedures, subject selection and exclusion criteria and the parameters to be used to monitor subject safety and assess efficacy. Each protocol, and any subsequent amendments to the protocol, must be submitted to the FDA as part of the IND. Furthermore, each clinical trial must be reviewed and approved by an IRB for each institution at which the clinical trial will be conducted to ensure that the risks to individuals participating in the clinical trials are minimized and are reasonable in relation to anticipated benefits. The IRB also approves the informed consent form that must be provided to each clinical trial subject or his or her legal representative, and must monitor the clinical trial until completed. There also are requirements governing the reporting of ongoing clinical trials and completed clinical trial results to public registries. Information about most clinical trials must be submitted within specific timeframes for publication on the *www.clinicaltrials.gov* website. Information related to the product, patient population, phase of investigation, study sites and investigators and other aspects of the clinical trial is made public as part of the registration of the clinical trial. Sponsors are also obligated to discuss the results of their clinical trials after completion. Disclosure of the results of these trials can be delayed in some cases for up to two years after the date of completion of the trial. Competitors may use this publicly available information to gain knowledge regarding the progress of development programs.

Human clinical trials are typically conducted in three sequential phases, which may overlap or be combined:

- Phase 1 clinical trials generally involve a small number of healthy volunteers or disease-affected patients who are initially exposed to a single dose and then multiple doses of the product candidate. The primary purpose of these clinical trials is to assess the metabolism, pharmacologic action, side effect tolerability and safety of the drug.

- Phase 2 clinical trials involve studies in disease-affected patients to determine the dose required to produce the desired benefits. At the same time, safety and further pharmacokinetic and pharmacodynamic information is collected, possible adverse effects and safety risks are identified and a preliminary evaluation of efficacy is conducted.

- Phase 3 clinical trials generally involve a larger number of patients at multiple sites and are designed to provide the data necessary to demonstrate the effectiveness of the product for its intended use, its safety in use and to establish the overall benefit/risk relationship of the product and provide an adequate basis for product approval. These trials may include comparisons with placebo and/or other comparator treatments. The duration of treatment is often extended to mimic the actual use of a product during marketing.

Post-approval trials, sometimes referred to as Phase 4 clinical trials, may be conducted after initial marketing approval. These trials are used to gain additional experience from the treatment of patients in the intended therapeutic indication, particularly for long-term safety follow up. In certain instances, the FDA may mandate the performance of Phase 4 clinical trials as a condition of approval of a BLA.

Progress reports detailing the results of the clinical trials must be submitted at least annually to the FDA and more frequently if serious adverse events occur. The FDA or the sponsor may suspend or terminate a clinical trial at any time or the FDA may impose other sanctions on various grounds, including a finding that the research patients are being exposed to an unacceptable health risk. Similarly, an IRB can refuse, suspend or terminate approval of a clinical trial at its institution if the clinical trial is not being conducted in accordance with the IRBs requirements or if the drug has been associated with unexpected serious harm to patients.

Concurrently with clinical trials, companies usually complete additional pre-clinical studies and must also develop additional information about the physical characteristics of the biological product as well as finalize a process for manufacturing the product in commercial quantities in accordance with cGMP requirements. To help reduce the risk of the introduction of adventitious agents with use of biological products, the PHSA emphasizes the importance of manufacturing control for products whose attributes cannot be precisely defined. The manufacturing process must be capable of consistently producing quality batches of the product candidate and, among other things, the sponsor must develop methods for testing the identity, strength, quality, potency and purity of the final biological product. Additionally, appropriate packaging must be selected and tested and stability studies must be conducted to demonstrate that the biological product candidate does not undergo unacceptable deterioration over its shelf life.

BLA review and approval

Assuming successful completion of the required clinical testing, the results of the pre-clinical studies and clinical trials, together with detailed information relating to the product's chemistry, manufacture, controls and proposed labeling, among other things, are submitted to the FDA as part of a BLA requesting approval to market the product for one or more indications. The BLA must contain proof of safety, purity, potency

and efficacy and may include both negative and ambiguous results of pre-clinical studies and clinical trials as well as positive findings. Data may come from company-sponsored clinical trials intended to test the safety and efficacy of a product's use or from a number of alternative sources, including studies initiated by investigators. To support marketing approval, the data submitted must be sufficient in quality and quantity to establish the safety and efficacy of the investigational product to the satisfaction of the FDA. FDA approval of a BLA must be obtained before a biologic may be marketed in the United States.

In most cases, the submission of a BLA is subject to a substantial application user fee. Under the goals and policies agreed to by the FDA under the Prescription Drug User Fee Act, or PDUFA, for original BLAs, the FDA has ten months from the filing date in which to complete its initial review of a standard application and respond to the applicant, and six months from the filing date for an application with priority review. The FDA does not always meet its PDUFA goal dates, and the review process is often significantly extended by FDA requests for additional information or clarification. This review typically takes twelve months from the date the BLA is submitted to the FDA because the FDA has approximately two months to make a "filing" decision.

Before approving a BLA, the FDA will typically conduct a pre-approval inspection of the manufacturing facilities for the new product to determine whether the manufacturing processes and facilities comply with cGMPs. The FDA will not approve the product unless it determines that the manufacturing processes and facilities are in compliance with cGMP requirements and adequate to assure consistent production of the product within required specifications. The FDA also may inspect the sponsor and one or more clinical trial sites to assure compliance with GCP requirements and the integrity of the clinical data submitted to the FDA.

Additionally, the FDA may refer applications for novel biologic candidates which present difficult questions of safety or efficacy to an advisory committee, typically a panel that includes clinicians and other experts, for review, evaluation and a recommendation as to whether the application should be approved and under what conditions, if any. The FDA is not bound by recommendations of an advisory committee, but it considers such recommendations when making decisions on approval. The FDA likely will re-analyze the clinical trial data, which could result in extensive discussions between the FDA and the applicant during the review process. The FDA also may require submission of a REMS plan if it determines that a REMS is necessary to ensure that the benefits of the drug outweigh its risks and to assure the safe use of the biological product. The REMS plan could include medication guides, physician communication plans, assessment plans and/or elements to assure safe use, such as restricted distribution methods, patient registries or other risk minimization tools. The FDA determines the requirement for a REMS, as well as the specific REMS provisions, on a case-by-case basis. If the FDA concludes a REMS is needed, the sponsor of the BLA must submit a proposed REMS. The FDA will not approve a BLA without a REMS, if required.

Under the Pediatric Research Equity Act, or PREA, a BLA or supplement to a BLA must contain data that are adequate to assess the safety and efficacy of the product candidate for the claimed indications in all relevant pediatric populations and to support dosing and administration for each pediatric population for which the product is safe and effective. The FDA may grant deferrals for submission of pediatric data or full or partial waivers. The Food and Drug Administration Safety and Innovation Act, or FDASIA, amended the FDCA to require that a sponsor who is planning to submit a marketing application for a product that includes a new active ingredient, new indication, new dosage form, new dosing regimen or new route of administration submit an initial Pediatric Study Plan, or PSP, within sixty days of an end-of-Phase 2 meeting or, if there is no such meeting, as early as practicable before the initiation of the Phase 3 or Phase 2/3 clinical trial. The initial PSP must include an outline of the pediatric study or studies that the sponsor plans to conduct, including trial objectives and design, age groups, relevant endpoints and

statistical approach, or a justification for not including such detailed information, and any request for a deferral of pediatric assessments or a full or partial waiver of the requirement to provide data from pediatric studies along with supporting information. The FDA and the sponsor must reach an agreement on the PSP. A sponsor can submit amendments to an agreed upon initial PSP at any time if changes to the pediatric plan need to be considered based on data collected from pre-clinical studies, early phase clinical trials or other clinical development programs.

After the FDA evaluates a BLA, it will issue an approval letter or a Complete Response Letter. An approval letter authorizes commercial marketing of the drug with specific prescribing information for specific indications. A Complete Response Letter indicates that the review cycle of the application is complete and the application will not be approved in its present form. A Complete Response Letter usually describes all of the specific deficiencies in the BLA identified by the FDA. The Complete Response Letter may require additional clinical or other data, additional pivotal Phase 3 clinical trial(s) and/or other significant and time-consuming requirements related to clinical trials, pre-clinical studies or manufacturing. If a Complete Response Letter is issued, the applicant may either resubmit the BLA, addressing all of the deficiencies identified in the letter, or withdraw the application. Even if such data and information are submitted, the FDA may decide that the re-submitted BLA does not satisfy the criteria for approval.

If a product receives regulatory approval, the approval is limited to the conditions of use (e.g., patient population, indication) described in the application. Further, depending on the specific risk(s) to be addressed, the FDA may require that contraindications, warnings or precautions be included in the product labeling, require that post-approval trials, including Phase 4 clinical trials, be conducted to further assess a product's safety after approval, require testing and surveillance programs to monitor the product after commercialization, or impose other conditions, including distribution and use restrictions or other risk management mechanisms under a REMS, which can materially affect the potential market and profitability of the product. The FDA may prevent or limit further marketing of a product based on the results of post-marketing trials or surveillance programs. After approval, some types of changes to the approved product, such as adding new indications, manufacturing changes and additional labeling claims, are subject to further testing requirements and FDA review and approval.

Orphan drug designation

Under the Orphan Drug Act, the FDA may grant orphan designation to a drug or biologic product intended to treat a rare disease or condition, which is generally a disease or condition that affects fewer than 200,000 individuals in the United States, or more than 200,000 individuals in the United States and for which there is no reasonable expectation that the cost of developing and making the product available in the United States for this type of disease or condition will be recovered from sales of the product in the United States. Orphan drug designation must be requested before submitting a BLA. After the FDA grants orphan drug designation, the identity of the therapeutic agent and its potential orphan use are disclosed publicly by the FDA. Orphan drug designation does not convey any advantage in, or shorten the duration of, the regulatory review and approval process. Orphan drug designation entitles a party to financial incentives such as opportunities for grant funding towards clinical trial costs, tax advantages and user-fee waivers. If a product that has orphan designation subsequently receives the first FDA approval for the disease or condition for which it has such designation, the product is entitled to orphan drug exclusivity, which means that the FDA may not approve any other applications to market the same drug for the same indication for seven years from the date of such approval, except in limited circumstances, such as a showing of clinical superiority to the product with orphan exclusivity by means of greater effectiveness, greater safety, by providing a major contribution to patient care or in instances of drug supply issues. Competitors, however, may receive approval of either a different product for the same indication or the

same product for a different indication that could be used "off-label" by physicians in the orphan indication, even though the competitor's product is not approved in the orphan indication. Orphan drug exclusivity also could block the approval of one of our products for seven years if a competitor obtains approval before we do of the same product, as defined by the FDA, for the same indication we are seeking, or if our product candidate is determined to be contained within the scope of the competitor's product for the same indication or disease. If one of our products designated as an orphan drug receives marketing approval for an indication broader than that which is designated, it may not be entitled to orphan drug exclusivity. Orphan drug status in the European Union has similar, but not identical, requirements and benefits.

Expedited review and approval

The FDA has various programs, including fast track designation, breakthrough therapy designation, accelerated approval and priority review, which are intended to expedite or simplify the process for the development and FDA review of drugs and biologics that are intended for the treatment of serious or life-threatening diseases or conditions and demonstrate the potential to address unmet medical needs. The purpose of these programs is to provide important new drugs and biologics to patients earlier than under standard FDA review procedures.

To be eligible for a fast track designation, the FDA must determine, based on the request of a sponsor, that a product is intended to treat a serious or life-threatening disease or condition and demonstrates the potential to address an unmet medical need by providing a therapy where none exists or a therapy that may be potentially superior to existing therapy based on efficacy or safety factors. Fast track designation provides opportunities for more frequent interactions with the FDA review team to expedite development and review of the product. The FDA may also review sections of the BLA for a fast track product on a rolling basis before the complete application is submitted, if the sponsor and the FDA agree on a schedule for the submission of the application sections, and the sponsor pays any required user fees upon submission of the first section of the BLA.

In addition, under the provisions of the or FDASIA, passed in July 2012, a sponsor can request designation of a product candidate as a "breakthrough therapy." A breakthrough therapy is defined as a drug or biologic that is intended, alone or in combination with one or more other drugs or biologics, to treat a serious or life-threatening disease or condition, and preliminary clinical evidence indicates that the drug or biologic may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. Drugs or biologics designated as breakthrough therapies are also eligible for accelerated approval. The FDA must take certain actions with respect to breakthrough therapies, such as holding timely meetings with and providing advice to the product sponsor, intended to expedite the development and review of an application for approval of a breakthrough therapy.

Products studied for their safety and effectiveness in treating serious or life-threatening illnesses and that provide meaningful therapeutic benefit over existing treatments may be eligible for accelerated approval and may be approved on the basis of adequate and well-controlled clinical trials establishing that the product has an effect on a surrogate endpoint that is reasonably likely to predict clinical benefit, or on a clinical endpoint that can be measured earlier than IMM, that is reasonably likely to predict an effect on IMM or other clinical benefit, taking into account the severity, rarity or prevalence of the condition and the availability or lack of alternative treatments. As a condition of approval, the FDA may require a sponsor of a product receiving accelerated approval to perform post-marketing trials to verify and describe the

predicted effect on IMM or other clinical endpoint, and the product may be subject to expedited withdrawal procedures.

Once a BLA is submitted for a product intended to treat a serious condition, the FDA may assign a priority review designation if the FDA determines that the product, if approved, would provide a significant improvement in safety or effectiveness. Under priority review, the FDA must review an application in six months, compared to ten months for a standard review. Most products that are eligible for fast track or breakthrough therapy designation are also likely to be considered appropriate to receive a priority review.

Even if a product qualifies for one or more of these programs, the FDA may later decide that the product no longer meets the conditions for qualification or decide that the time period for FDA review or approval will not be shortened. Furthermore, fast track designation, breakthrough therapy designation, accelerated approval and priority review do not change the standards for approval and may not ultimately expedite the development or approval process.

Post-approval requirements

Following approval of a new product, the manufacturer and the approved product are subject to pervasive and continuing regulation by the FDA, including, among other things, monitoring and recordkeeping activities, reporting of adverse experiences with the product, product sampling and distribution restrictions, complying with promotion and advertising requirements, which include restrictions on promoting drugs for unapproved uses or patient populations (i.e., "off-label use") and limitations on industry-sponsored scientific and educational activities. Although physicians may prescribe legally available products for off-label uses, manufacturers may not market or promote such uses. The FDA and other agencies actively enforce the laws and regulations prohibiting the promotion of off-label uses, and a company that is found to have improperly promoted off-label uses may be subject to significant liability. If there are any modifications to the product, including changes in indications, labeling or manufacturing processes or facilities, the applicant may be required to submit and obtain FDA approval of a new BLA or BLA supplement, which may require the applicant to develop additional data or conduct additional pre-clinical studies and clinical trials. The FDA may also place other conditions on approvals including the requirement for a REMS to assure the safe use of the product. A REMS could include medication guides, physician communication plans or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools. Any of these limitations on approval or marketing could restrict the commercial promotion, distribution, prescription or dispensing of products. Product approvals may be withdrawn for non-compliance with regulatory standards or if problems occur following initial marketing.

FDA regulations require that products be manufactured in specific approved facilities and in accordance with cGMPs. We rely, and expect to continue to rely, on third parties for the production of clinical and commercial quantities of our products in accordance with cGMP regulations. These manufacturers must comply with cGMP regulations that require, among other things, quality control and quality assurance, the maintenance of records and documentation and the obligation to investigate and correct any deviations from cGMP. Manufacturers and other entities involved in the manufacture and distribution of approved drugs or biologics are required to register their establishments with the FDA and certain state agencies, and are subject to periodic unannounced inspections by the FDA and certain state agencies for compliance with cGMP and other laws. Accordingly, manufacturers must continue to expend time, money and effort in the area of production and quality control to maintain cGMP compliance. The discovery of conditions that violate these rules, including failure to conform to cGMPs, could result in enforcement actions, and the

discovery of problems with a product after approval may result in restrictions on a product, manufacturer or holder of an approved BLA, including voluntary recall.

Once an approval or clearance of a drug is granted, the FDA may withdraw the approval if compliance with regulatory requirements and standards is not maintained or if problems occur after the product reaches the market. Later discovery of previously unknown problems with a product, including adverse events of unanticipated severity or frequency, or with manufacturing processes, or failure to comply with regulatory requirements, may result in mandatory revisions to the approved labeling to add new safety information; imposition of post-market or clinical trials to assess new safety risks; or imposition of distribution or other restrictions under a REMS program. Other potential consequences include, among other things:

- restrictions on the marketing or manufacturing of the product, complete withdrawal of the product from the market or product recalls;
- fines, warning letters or other enforcement-related letters or clinical holds on post-approval clinical trials;
- refusal of the FDA to approve pending BLAs or supplements to approved BLAs, or suspension or revocation of product approvals;
- product seizure or detention, or refusal to permit the import or export of products;
- injunctions or the imposition of civil or criminal penalties; and
- consent decrees, corporate integrity agreements, debarment, or exclusion from federal healthcare programs; or mandated modification of promotional materials and labeling and the issuance of corrective information.

Biosimilars and exclusivity

An abbreviated approval pathway for biological products that are biosimilar to or interchangeable with an FDA-licensed reference biological product was created by the Biologics Price Competition and Innovation Act of 2009, or BPCIA, as part of the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act, or the Affordable Care Act. This amendment to the PHSA, in part, attempts to minimize duplicative testing. 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 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. To date, the FDA has approved a number of biosimilars, and numerous biosimilars have been approved in Europe. The FDA has issued several guidance documents outlining its approach to reviewing and approving biosimilars.

Biosimilarity, which requires that the biological product be highly similar to the reference product notwithstanding minor differences in clinically inactive components and that there be no clinically meaningful differences between the product and the reference product in terms of safety, purity and potency, must be shown through analytical studies, animal studies and a clinical trial or trials. Interchangeability requires that a biological product be biosimilar to the reference product and that the product can be expected to produce the same clinical results as the reference product in any given patient and, for products administered multiple times to an individual, that the product and the reference product may be alternated or switched after one has been previously administered without increasing safety risks

or risks of diminished efficacy relative to exclusive use of the reference biological product without such alternation or switch. Complexities associated with the larger, and often more complex, structure of biological products as compared to small molecule drugs, as well as the processes by which such products are manufactured, pose significant hurdles to implementation that are still being worked out by the FDA.

A reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, and the FDA will not accept an application for a biosimilar or interchangeable product based on the reference biological product until four years after the date of first licensure of the reference product. "First licensure" typically means the initial date the particular product at issue was licensed in the United States. Date of first licensure does not include the date of licensure of (and a new period of exclusivity is not available for) a supplement for the reference product for a subsequent application filed by the same sponsor or manufacturer of the reference product (or licensor, predecessor in interest or other related entity) for a change (not including a modification to the structure of the biological product) that results in a new indication, route of administration, dosing schedule, dosage form, delivery system, delivery device or strength or for a modification to the structure of the biological product that does not result in a change in safety, purity or potency. Therefore, one must determine whether a new product includes a modification to the structure of a previously licensed product that results in a change in safety, purity or potency to assess whether the licensure of the new product is a first licensure that triggers its own period of exclusivity. Whether a subsequent application, if approved, warrants exclusivity as the "first licensure" of a biological product is determined on a case-by-case basis with data submitted by the sponsor.

The BPCIA is complex and only beginning to be interpreted and implemented by the FDA. In addition, recent government proposals have sought to reduce the 12-year reference product exclusivity period. Other aspects of the BPCIA, some of which may impact the BPCIA exclusivity provisions, have also been the subject of recent litigation. As a result, the ultimate impact, implementation and meaning of the BPCIA is subject to significant uncertainty.

U.S. patent term restoration

Depending upon the timing, duration and conditions of FDA marketing approval of our product candidates, one or more of our U.S. patents may be eligible for limited patent term extension under the Drug Price Competition and Patent Term Restoration Act of 1984, referred to as the Hatch-Waxman Amendments, and similar legislation in the European Union. The Hatch-Waxman Amendments permit a patent term extension of up to five years for a patent covering an approved product as compensation for effective patent term lost during product development and the FDA regulatory review process. However, we may not receive an extension if we fail to apply within applicable deadlines, fail to apply prior to expiration of relevant patents or otherwise fail to satisfy applicable requirements. Moreover, the length of the extension could be less than we request. Only one patent per approved product can be extended, the extension must be based on the first approval for the product, and the extension cannot extend the total patent term beyond fourteen years from approval. If we are unable to obtain patent term extension or the term of any such extension is less than we request, the period during which we can enforce our patent rights for that product will be shortened and our competitors may obtain approval to market competing products sooner.

European union drug development, review and approval

In the European Union, our product candidates also may be subject to extensive regulatory requirements. As in the United States, medicinal products can be marketed only if a marketing authorization from the competent regulatory agencies has been obtained.

Similar to the United States, the various phases of pre-clinical and clinical research in the European Union are subject to significant regulatory controls.

The Clinical Trials Directive 2001/20/EC, the Directive 2005/28/EC on GCP, and the related national implementing provisions of the individual EU Member States govern the system for the approval of clinical trials in the European Union. Under this system, an applicant must obtain prior approval from the competent national authority of the EU Member States in which the clinical trial is to be conducted. Furthermore, the applicant may only start a clinical trial at a specific study site after the competent ethics committee has issued a favorable opinion. The clinical trial application must be accompanied by, among other documents, an IMPD (the Common Technical Document) with supporting information prescribed by Directive 2001/20/EC, Directive 2005/28/EC, where relevant the implementing national provisions of the individual EU Member States and further detailed in applicable guidance documents. All suspected unexpected serious adverse reactions to the investigated drug that occur during the clinical trial have to be reported to the competent national authority and the Ethics Committee of the Member State where they occurred.

In April 2014, the new Clinical Trials Regulation, (EU) No 536/2014 (Clinical Trials Regulation) was adopted. The Regulation is anticipated to come into application in 2019. The Clinical Trials Regulation will be directly applicable in all the EU Member States, repealing the current Clinical Trials Directive 2001/20/EC. Conduct of all clinical trials performed in the European Union will continue to be bound by currently applicable provisions until the new Clinical Trials Regulation becomes applicable. The extent to which ongoing clinical trials will be governed by the Clinical Trials Regulation will depend on when the Clinical Trials Regulation becomes applicable and on the duration of the individual clinical trial. If a clinical trial continues for more than three years from the day on which the Clinical Trials Regulation becomes applicable the Clinical Trials Regulation will at that time begin to apply to the clinical trial.

The new Clinical Trials Regulation aims to simplify and streamline the approval of clinical trials in the European Union. The main characteristics of the regulation include: a streamlined application procedure via a single entry point, the "EU portal"; a single set of documents to be prepared and submitted for the application as well as simplified reporting procedures for clinical trial sponsors; and a harmonized procedure for the assessment of applications for clinical trials, which is divided in two parts. Part I is assessed by the competent authorities of all EU Member States in which an application for authorization of a clinical trial has been submitted (Member States concerned). Part II is assessed separately by each Member State concerned. Strict deadlines have been established for the assessment of clinical trial applications. The role of the relevant ethics committees in the assessment procedure will continue to be governed by the national law of the concerned EU Member State. However, overall related timelines will be defined by the Clinical Trials Regulation.

To obtain a marketing authorization of a drug in the European Union, we may submit marketing authorization applications, or MAA, either under the so-called centralized or national authorization procedures.

Centralized procedure

The centralized procedure provides for the grant of a single marketing authorization following a favorable opinion by the European Medicines Agency, or EMA, that is valid in all EU member states, as well as Iceland, Liechtenstein and Norway. The centralized procedure is compulsory for medicines produced by specified biotechnological processes, products designated as orphan medicinal products, advanced-therapy medicines (such as gene-therapy, somatic cell-therapy or tissue-engineered medicines) and products with a new active substance indicated for the treatment of specified diseases, such as HIV/AIDS, cancer, diabetes,

neurodegenerative disorders or autoimmune diseases and other immune dysfunctions and viral diseases. The centralized procedure is optional for products that represent a significant therapeutic, scientific or technical innovation, or whose authorization would be in the interest of public health. Under the centralized procedure the maximum timeframe for the evaluation of an MAA by the EMA is 210 days, excluding clock stops, when additional written or oral information is to be provided by the applicant in response to questions asked by the Committee of Medicinal Products for Human Use, or the CHMP. Accelerated assessment might be granted by the CHMP in exceptional cases, when a medicinal product is expected to be of a major public health interest, particularly from the point of view of therapeutic innovation. The timeframe for the evaluation of an MAA under the accelerated assessment procedure is of 150 days, excluding stop-clocks.

National authorization procedures

There are also two other possible routes to authorize medicinal products in several EU countries, which are available for investigational medicinal products that fall outside the scope of the centralized procedure:

- Decentralized procedure. Using the decentralized procedure, an applicant may apply for simultaneous authorization in more than one EU country of medicinal products that have not yet been authorized in any EU country and that do not fall within the mandatory scope of the centralized procedure.

- Mutual recognition procedure. In the mutual recognition procedure, a medicine is first authorized in one EU Member State, in accordance with the national procedures of that country. Following this, further marketing authorizations can be sought from other EU countries in a procedure whereby the countries concerned agree to recognize the validity of the original, national marketing authorization.

Under the above described procedures, before granting the MA, the EMA or the competent authorities of the Member States of the EEA make an assessment of the risk-benefit balance of the product on the basis of scientific criteria concerning its quality, safety and efficacy.

European union regulatory exclusivity

In the European Union, new products authorized for marketing (i.e., reference products) qualify for eight years of data exclusivity and an additional two years of market exclusivity upon marketing authorization. The data exclusivity period prevents generic applicants from relying on the pre-clinical and clinical trial data contained in the dossier of the reference product when applying for a generic marketing authorization in the European Union during a period of eight years from the date on which the reference product was first authorized in the European Union. The market exclusivity period prevents a successful generic applicant from commercializing its product in the EU until ten years have elapsed from the initial authorization of the reference product in the EU. The ten-year market exclusivity period can be extended to a maximum of eleven years if, during the first eight years of those ten years, the marketing authorization holder obtains an authorization for one or more new therapeutic indications which, during the scientific evaluation prior to their authorization, are held to bring a significant clinical benefit in comparison with existing therapies.

European union orphan designation and exclusivity

The criteria for designating an orphan medicinal product in the European Union, are similar in principle to those in the United States. Under Article 3 of Regulation (EC) 141/2000, a medicinal product may be designated as orphan if (1) it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition; (2) either (a) such condition affects no more than five in 10,000 persons in the European Union when the application is made, or (b) the product, without the benefits derived from

orphan status, would not generate sufficient return in the European Union to justify investment; and (3) there exists no satisfactory method of diagnosis, prevention or treatment of such condition authorized for marketing in the European Union, or if such a method exists, the product will be of significant benefit to those affected by the condition, as defined in Regulation (EC) 847/2000. Orphan medicinal products are eligible for financial incentives such as reduction of fees or fee waivers and are, upon grant of a marketing authorization, entitled to ten years of market exclusivity for the approved therapeutic indication. The application for orphan designation must be submitted before the application for marketing authorization. The applicant will receive a fee reduction for the marketing authorization application if the orphan designation has been granted, but not if the designation is still pending at the time the marketing authorization is submitted. Orphan designation does not convey any advantage in, or shorten the duration of, the regulatory review and approval process.

The ten-year market exclusivity in the European Union may be reduced to six years if, at the end of the fifth year, it is established that the product no longer meets the criteria for orphan designation, for example, if the product is sufficiently profitable not to justify maintenance of market exclusivity. Additionally, marketing authorization may be granted to a similar product for the same indication at any time if:

- the second applicant can establish that its product, although similar, is safer, more effective or otherwise clinically superior;
- the applicant consents to a second orphan medicinal product application; or
- the applicant cannot supply enough orphan medicinal product.

Rest of the world regulation

For other countries outside of the European Union and the United States, such as countries in Eastern Europe, Latin America or Asia, the requirements governing the conduct of clinical trials, product licensing, pricing and reimbursement vary from jurisdiction to jurisdiction. Additionally, the clinical trials must be conducted in accordance with cGCP requirements and the applicable regulatory requirements and the ethical principles that have their origin in the Declaration of Helsinki.

If we fail to comply with applicable foreign regulatory requirements, we may be subject to, among other things, fines, suspension or withdrawal of regulatory approvals, product recalls, seizure of products, operating restrictions and criminal prosecution.

Other healthcare laws

In addition to FDA restrictions on the marketing of pharmaceutical products, other U.S., federal and state healthcare regulatory laws restrict business practices in the pharmaceutical industry. These laws include, but are not limited to, federal and state anti-kickback, false claims, data privacy and security and physician payment and drug pricing transparency laws.

The U.S. federal Anti-Kickback Statute prohibits, among other things, any person or entity from knowingly and willfully offering, paying, soliciting, receiving or providing any remuneration, directly or indirectly, overtly or covertly, to induce or in return for purchasing, leasing, ordering or arranging for or recommending the purchase, lease or order of any good, facility, item or service reimbursable, in whole or in part, under Medicare, Medicaid or other federal healthcare programs. The term "remuneration" has been broadly interpreted to include anything of value. The Anti-Kickback Statute has been interpreted to apply to arrangements between pharmaceutical and medical device manufacturers on the one hand and

prescribers, purchasers, formulary managers and beneficiaries on the other hand. Although there are a number of statutory exceptions and regulatory safe harbors protecting some common activities from prosecution, the exceptions and safe harbors are drawn narrowly. Practices that involve remuneration that may be alleged to be intended to induce prescribing, purchases or recommendations may be subject to scrutiny if they do not meet the requirements of a statutory or regulatory exception or safe harbor. Failure to meet all of the requirements of a particular applicable statutory exception or regulatory safe harbor does not make the conduct per se illegal under the U.S. federal Anti-Kickback Statute. Instead, the legality of the arrangement will be evaluated on a case-by-case basis based on a cumulative review of all its facts and circumstances. Several courts have interpreted the statute's intent requirement to mean that if any one purpose of an arrangement involving remuneration is to induce referrals of federal healthcare covered business, the statute has been violated. In addition, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation. Moreover, a claim including items or services resulting from a violation of the U.S. federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the federal civil False Claims Act. The majority of states also have anti-kickback laws, which establish similar prohibitions and in some cases may apply to items or services reimbursed by any third-party payor, including commercial insurers, or to self-pay patients.

The federal false claims and civil monetary penalties laws, including the civil False Claims Act, prohibit, among other things, any person or entity from knowingly presenting, or causing to be presented, a false, fictitious or fraudulent claim for payment to, or approval by, the federal government, knowingly making, using or causing to be made or used a false record or statement material to a false or fraudulent claim to the federal government, or knowingly making a false statement to avoid, decrease or conceal an obligation to pay money to the U.S. federal government. A claim includes "any request or demand" for money or property presented to the U.S. government. Actions under the civil False Claims Act may be brought by the Attorney General or as a *qui tam* action by a private individual in the name of the government. Violations of the civil False Claims Act can result in very significant monetary penalties and treble damages. Several biopharmaceutical, medical device and other healthcare companies have been prosecuted under these laws for, among other things, allegedly providing free product to customers with the expectation that the customers would bill federal programs for the product. Other companies have been prosecuted for causing false claims to be submitted because of the companies' marketing of products for unapproved (e.g., or off-label), and thus non-covered, uses. In addition, the civil monetary penalties statute imposes penalties against any person who is determined to have presented or caused to be presented a claim to a federal health program that the person knows or should know is for an item or service that was not provided as claimed or is false or fraudulent. Many states also have similar fraud and abuse statutes or regulations that apply to items and services reimbursed under Medicaid and other state programs, or, in several states, apply regardless of the payor.

Violations of fraud and abuse laws, including federal and state anti-kickback and false claims laws, may be punishable by criminal and civil sanctions, including fines and civil monetary penalties, the possibility of exclusion from federal healthcare programs (including Medicare and Medicaid), disgorgement of profits and corporate integrity agreements, which impose, among other things, rigorous operational and monitoring requirements on companies. Similar sanctions and penalties, as well as imprisonment, also can be imposed upon executive officers and employees of such companies. Given the significant size of actual and potential settlements, it is expected that the government authorities will continue to devote substantial resources to investigating healthcare providers' and manufacturers' compliance with applicable fraud and abuse laws.

The federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, created additional federal criminal statutes that prohibit, among other actions, knowingly and willfully executing, or attempting to execute, a scheme to defraud any healthcare benefit program, including private third-party payors, knowingly and willfully embezzling or stealing from a healthcare benefit program, willfully obstructing a criminal investigation of a healthcare offense and knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false, fictitious or fraudulent statement in connection with the delivery of or payment for healthcare benefits, items or services. Similar to the U.S. federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation.

In addition, there has been a recent trend of increased federal and state regulation of payments made to physicians and certain other healthcare providers. The Affordable Care Act imposed, among other things, new annual reporting requirements through the Physician Payments Sunshine Act for covered manufacturers for certain payments and "transfers of value" provided to physicians and teaching hospitals, as well as ownership and investment interests held by physicians and their immediate family members. Failure to submit timely, accurately and completely the required information for all payments, transfers of value and ownership or investment interests may result in civil monetary penalties. Covered manufacturers must submit reports by the 90th day of each subsequent calendar year and the reported information is publicly made available on a searchable website. In addition, certain states require implementation of compliance programs and compliance with the pharmaceutical industry's voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government, impose restrictions on marketing practices and/or require the tracking and reporting of marketing expenditures and pricing information as well as gifts, compensation and other remuneration or items of value provided to physicians and other healthcare professionals and entities.

We may also be subject to data privacy and security regulation by both the federal government and the states in which we conduct our business. HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act, or HITECH, and their respective implementing regulations, including the Final HIPAA Omnibus Rule published on January 25, 2013, impose specified requirements relating to the privacy, security and transmission of individually identifiable health information held by covered entities and their business associates. Among other things, HITECH made HIPAAs security standards directly applicable to "business associates," defined as independent contractors or agents of covered entities that create, receive, maintain or transmit protected health information in connection with providing a service for or on behalf of a covered entity, although it is unclear that we would be considered a "business associate" in the normal course of our business. HITECH also increased the civil and criminal penalties that may be imposed against covered entities, business associates and possibly other persons, and gave state attorneys general new authority to file civil actions for damages or injunctions in federal courts to enforce the federal HIPAA laws and seek attorney's fees and costs associated with pursuing federal civil actions. In addition, state laws govern the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and may not have the same requirements, thus complicating compliance efforts.

Similar foreign laws and regulations, which may include, for instance, applicable post-marketing requirements, anti-fraud and abuse laws and implementation of corporate compliance programs and reporting of payments or other transfers of value to healthcare professionals, may apply to us to the extent that any of our product candidates, once approved, are sold in a country other than the United States.

Because of the breadth of these laws and the narrowness of their exceptions and safe harbors, it is possible that business activities can be subject to challenge under one or more of such laws. The scope and enforcement of each of these laws is uncertain and subject to rapid change in the current environment of healthcare reform, especially in light of the lack of applicable precedent and regulations. Federal and state enforcement bodies have recently increased their scrutiny of interactions between healthcare companies and healthcare providers, which has led to a number of investigations, prosecutions, convictions and settlements in the healthcare industry.

Ensuring that business arrangements with third parties comply with applicable healthcare laws and regulations is costly and time consuming. If business operations are found to be in violation of any of the laws described above or any other applicable governmental regulations a pharmaceutical manufacturer may be subject to penalties, including civil, criminal and administrative penalties, damages, fines, disgorgement of profits, individual imprisonment, exclusion from governmental funded healthcare programs, such as Medicare and Medicaid, contractual damages, reputational harm, diminished profits and future earnings, additional reporting obligations and oversight if subject to a corporate integrity agreement or other agreement to resolve allegations of non-compliance with these laws, and curtailment or restructuring of operations, any of which could adversely affect a pharmaceutical manufacturer's ability to operate its business and the results of its operations.

Coverage, pricing and reimbursement

Significant uncertainty exists as to the coverage and reimbursement status of any biological products for which we obtain regulatory approval. The United States government, state legislatures and foreign governments have shown significant interest in implementing cost containment programs to limit the growth of government-paid healthcare costs, including price-controls, restrictions on reimbursement and requirements for substitution of generic products for branded drug and biologic products. In the United States and markets in other countries, patients who are prescribed products generally rely on third-party payors to reimburse all or part of the associated healthcare costs. Providers and patients are unlikely to use our products unless coverage is provided and reimbursement is adequate to cover a significant portion of the cost of our products. If approved, sales of our product candidates will depend, in part, on the availability of coverage and adequate reimbursement from third-party payors. Third-party payors include government authorities, managed care plans, private health insurers and other organizations.

In the United States, the process for determining whether a third-party payor will provide coverage for a biological product typically is separate from the process for setting the price of such product or for establishing the reimbursement rate that the payor will pay for the product once coverage is approved. With respect to biologics, third-party payors may limit coverage to specific products on an approved list, also known as a formulary, which might not include all of the FDA-approved products for a particular indication, or place products at certain formulary levels that result in lower reimbursement levels and higher cost-sharing obligation imposed on patients. A decision by a third-party payor not to cover our product candidates could reduce physician utilization of a product. Moreover, a third-party payor's decision to provide coverage for a product does not imply that an adequate reimbursement rate will be approved. Adequate third-party reimbursement may not be available to enable a manufacturer to maintain price levels sufficient to realize an appropriate return on its investment in product development. Additionally, coverage and reimbursement for products can differ significantly from payor to payor. One third-party payor's decision to cover a particular medical product does not ensure that other payors will also provide coverage for the medical product, or will provide coverage at an adequate reimbursement rate. As a result, the coverage determination process usually requires manufacturers to provide scientific and clinical support for the use of their products to each payor separately and is a time-consuming process.

As noted above, the marketability of any products for which we receive regulatory approval for commercial sale may suffer if the government and third-party payors fail to provide coverage and adequate reimbursement. An increasing emphasis on cost containment measures in the United States has increased and we expect will continue to increase the pressure on pharmaceutical pricing. Coverage policies and third-party reimbursement rates may change at any time. Even if favorable coverage and reimbursement status is attained for one or more products for which we receive regulatory approval, less favorable coverage policies and reimbursement rates may be implemented in the future. Third-party payors are increasingly challenging the prices charged for medical products and services, examining the medical necessity and reviewing the cost-effectiveness of pharmaceutical products, in addition to questioning safety and efficacy. If third-party payors do not consider a product to be cost-effective compared to other available therapies, they may not cover that product after FDA approval or, if they do, the level of payment may not be sufficient to allow a manufacturer to sell its product at a profit.

In addition, in many foreign countries, the proposed pricing for a drug must be approved before it may be lawfully marketed. The requirements governing drug pricing and reimbursement vary widely from country to country. In the European Union, governments influence the price of products through their pricing and reimbursement rules and control of national healthcare systems that fund a large part of the cost of those products to consumers. Some jurisdictions operate positive and negative list systems under which products may only be marketed once a reimbursement price has been agreed to by the government. To obtain reimbursement or pricing approval, some of these countries may require the completion of clinical trials that compare the cost effectiveness of a particular product to currently available therapies. Other member states allow companies to fix their own prices for medicines, but monitor and control company profits. There can be no assurance that any country that has price controls or reimbursement limitations for pharmaceutical products will allow favorable reimbursement and pricing arrangements for any of our products. The downward pressure on healthcare costs in general, particularly prescription products, has become very intense. As a result, increasingly high barriers are being erected to the entry of new products. In addition, in some countries, cross border imports from low-priced markets exert a commercial pressure on pricing within a country.

Healthcare reform and potential changes to healthcare laws

The FDAs and other regulatory authorities' policies may change and additional government regulations may be enacted that could prevent, limit or delay regulatory approval of our product candidates. For example, in December 2016, the 21st Century Cures Act, or Cures Act, was signed into law. The Cures Act, among other things, is intended to modernize the regulation of drugs and devices and to spur innovation, but its ultimate implementation is uncertain. In addition, in August 2017, the FDA Reauthorization Act was signed into law, which reauthorized the FDAs user fee programs and included additional drug and device provisions that build on the Cures Act. If we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we are not able to maintain regulatory compliance, we may lose any marketing approval that we otherwise may have obtained and we may not achieve or sustain profitability, which would adversely affect our business, prospects, financial condition and results of operations.

A primary trend in the U.S. healthcare industry and elsewhere is cost containment. Government authorities and other third-party payors have attempted to control costs by limiting coverage and the amount of reimbursement for particular medical products and services, implementing reductions in Medicare and other healthcare funding and applying new payment methodologies. For example, in March 2010, the Affordable Care Act was enacted, which, among other things, increased the minimum Medicaid rebates owed by most manufacturers under the Medicaid Drug Rebate Program; introduced a new methodology by

which rebates owed by manufacturers under the Medicaid Drug Rebate Program are calculated for drugs that are inhaled, infused, instilled, implanted or injected; extended the Medicaid Drug Rebate Program to utilization of prescriptions of individuals enrolled in Medicaid managed care plans; imposed mandatory discounts for certain Medicare Part D beneficiaries as a condition for manufacturers' outpatient drugs coverage under Medicare Part D; subjected drug manufacturers to new annual fees based on pharmaceutical companies' share of sales to federal healthcare programs; imposed a new federal excise tax on the sale of certain medical devices; created a new Patient Centered Outcomes Research Institute to oversee, identify priorities in, and conduct comparative clinical effectiveness research, along with funding for such research; created the Independent Payment Advisory Board, which, once empaneled, will have authority to recommend certain changes to the Medicare program that could result in reduced payments for prescription drugs; and established a Center for Medicare Innovation at the CMS to test innovative payment and service delivery models to lower Medicare and Medicaid spending.

Since its enactment, there have been judicial and Congressional challenges to certain aspects of the Affordable Care Act, and we expect there will be additional challenges and amendments to the Affordable Care Act in the future. The current Presidential administration and members of the U.S. Congress have indicated that they may continue to seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Most recently, the Tax Cuts and Jobs Acts was enacted, which, among other things, removes penalties for not complying with the individual mandate to carry health insurance. 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 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 to reduce healthcare expenditures. These changes include the Budget Control Act of 2011, which, among other things, included aggregate reductions of Medicare payments to providers of 2% per fiscal year and that will remain in effect through 2025 unless additional action is taken by Congress; and the American Taxpayer Relief Act of 2012, which, among other things, further reduced Medicare payments to several types of providers, including hospitals, imaging centers and cancer treatment centers and increased the statute of limitations period for the government to recover overpayments to providers from three to five years. More recently, there has been heightened governmental scrutiny over the manner in which manufacturers set prices for their marketed products, which has resulted in several Congressional inquiries and proposed and enacted federal and state legislation designed to, among other things, bring more transparency to product pricing, review the relationship between pricing and manufacturer patient programs and reform government program reimbursement methodologies for pharmaceutical and biologic products.

Individual states in the United States have become increasingly active in passing legislation and implementing regulations designed to control biotechnology and pharmaceutical product pricing, including price or patient reimbursement constraints, discounts, restrictions on certain product access and marketing cost disclosure and transparency measures, and, in some cases, designed to encourage importation from other countries and bulk purchasing.

We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative or executive action, either in the United States or abroad. We expect that

additional state and federal healthcare reform measures will be adopted in the future, any of which could limit the amounts that federal and state governments will pay for healthcare products and services.

Employees

As of December 31, 2018, we had 111 employees.

Facilities

Our U.S. headquarters are located in Lexington, Massachusetts, where Kiniksa US has leased approximately 55,924 square feet of office and laboratory space, under a lease which expires in July 2021. We believe that our offices are sufficient to meet our current needs and that suitable additional space will be available as and when needed.

Legal proceedings

We are not subject to any material legal proceedings.

Management

Executive officers and directors

The following table sets forth the name and position of each of our executive officers and directors and their ages as of the date of this prospectus.

Name	Age	Position
Executive Officers:		
Sanj K. Patel	49	Chief Executive Officer and Chairman of the Board
Stephen Mahoney	48	President and Chief Operating Officer
Chris Heberlig	44	Chief Financial Officer
John F. Paolini, M.D., Ph.D.	54	Chief Medical Officer
Thomas Beetham	49	Chief Legal Officer
Directors:		
Felix J. Baker, Ph.D.(2)(3)	49	Director
Stephen R. Biggar, M.D., Ph.D.(3)	48	Director
Thomas R. Malley(1)(3)	50	Director
Tracey L. McCain(1)	51	Director
Kimberly J. Popovits(2)	60	Director
Barry D. Quart, Pharm.D.(1)(2)	62	Director

(1) Member of the audit committee.

(2) Member of the compensation committee.

(3) Member of the nominating and corporate governance committee.

Executive officers

Sanj K. Patel has served as our Chief Executive Officer and Chairman of our board of directors since our formation in July 2015. In June 2008, Mr. Patel formed Synageva BioPharma Corp., or Synageva, a biotechnology company focused on rare diseases, where he acted as President and Chief Executive Officer until its sale to Alexion Pharmaceuticals in June 2015. Prior to Synageva, Mr. Patel held various roles at Genzyme Corporation from 1999 to 2008, most recently as head of U.S. Sales, Marketing and Commercial Operations for the Genzyme Therapeutics franchise, or Genzyme. Mr. Patel is a member of the board of directors of Syros Pharmaceuticals and BioCryst Pharmaceuticals, and, from 2013 to 2015, sat on the board of directors of Intercept Pharmaceuticals. He is also the founder and director of the Sanj K. Patel and Family Foundation, a philanthropic organization that supports charities for patients with rare and devastating diseases. Mr. Patel holds a B.Sc. with Honors from the University of the South Bank, London and completed his management and business studies at Ealing College, London and his Pharmacology research program at the Wellcome Foundation. We believe that Mr. Patel is qualified to serve on our board of directors due to his extensive business, sales and product development experience in the biotechnology industry.

Stephen Mahoney has served as our Chief Operating Officer since our formation in July 2015 and as our President since June 2017. Prior to serving as our Chief Operating Officer, Mr. Mahoney held various roles at Synageva from 2012 to 2015, most recently as Chief Commercial Officer, where he was responsible for Synageva's global commercial operations. Mr. Mahoney was also responsible for areas such as Global Sales Operations & Business Analytics, Commercial Supply Chain and Logistics, Global Procurement, Patient Services, Sales Training and Legal and Corporate Development. Prior to Synageva, Mr. Mahoney held

various roles at Genzyme from 2003 to 2012, most recently as the Regional Legal Director for the Asia Pacific region, where he was responsible for legal and healthcare compliance issues for multiple business units. Mr. Mahoney holds an M.B.A. from Boston College's Carroll School of Management, a J.D. from Boston College Law School and a B.A. from Colorado College.

Chris Heberlig has served as our Chief Financial Officer since our formation in July 2015. Prior to serving as our Chief Financial Officer, Mr. Heberlig held various roles at Synageva from 2008 to 2015, most recently serving as Senior Vice President of Finance and Business Operations. At Synageva, he led strategic tax planning, including overseeing the transfer of tax and intellectual property assets to Europe, and was responsible for global financial operations, facilities, as well as program management. Mr. Heberlig holds an M.B.A. from Boston University School of Management and a B.A. from St. Lawrence University. Mr. Heberlig is also a Certified Public Accountant.

John F. Paolini, *M.D., Ph.D.*, has served as our Chief Medical Officer since August 2016. From August 2015 to August 2016, Dr. Paolini was Clinical Research Head of the Cardiovascular and Metabolic Diseases Research Unit at Pfizer Inc., a pharmaceutical company, where he was responsible for bringing forward programs from pre-clinical through early clinical development and proof of concept. Prior to Pfizer, from August 2011 to July 2015, Dr. Paolini served as Chief Medical Officer of Cerenis Therapeutics, a biotechnology company focused on cardiovascular and metabolic diseases, where he was responsible for designing and executing clinical trials and regulatory strategy for a portfolio of products. Dr. Paolini holds an M.D., Ph.D. from Duke University School of Medicine and a B.A. and a Bachelor of Science, or B.S., from Tulane University, and completed his internship, residency and fellowship in Internal Medicine and Cardiology from Brigham and Women's Hospital, Boston.

Thomas Beetham has served as our Chief Legal Officer since our formation in July 2015 and is also responsible for corporate development. Prior to serving as our Chief Legal Officer, Mr. Beetham was the Chief Legal Officer and Senior Vice President of Corporate Development for Synageva from October 2013 to June 2015. At Synageva, in addition to leading the legal department, Mr. Beetham was responsible for business development activities. Prior to joining Synageva, from October 2011 to October 2013, Mr. Beetham was the General Legal Counsel for New England Biolabs, Inc., a reagent supplier for genomic research, where he was responsible for legal matters and a member of Biolabs' global business development team. Before Synageva, Mr. Beetham was at Genzyme from September 2004 to October 2011, most recently as the lead corporate attorney responsible for Genzyme's hematology/oncology and multiple sclerosis products, and from September 1999 to September 2004 was a corporate and transactional attorney with the law firm of Palmer & Dodge, LLP. Mr. Beetham holds an M.B.A. from Boston College's Carroll School of Management, a J.D. from Boston College Law School and a B.A. from the University of Rochester.

Directors

Felix J. Baker, Ph.D., has served as our lead director and on our board of directors since October 2015. Since 2000, Dr. Baker has been a Co-Managing Member of Baker Bros. Advisors LP, or Baker Brothers, an investment advisor focused on investments in life science and biotechnology companies. Dr. Baker and his brother, Julian Baker, started their fund management careers in 1994 when they co-founded a biotechnology investing partnership with the Tisch Family. Dr. Baker currently serves on the boards of directors of Alexion Pharmaceuticals, Genomic Health, Inc., Seattle Genetics, Inc., and Kodiak Sciences Inc. and previously served on the board of directors of Synageva. Dr. Baker holds a B.S. and a Ph.D. in Immunology from Stanford University, where he also completed two years of medical school. We believe Dr. Baker is qualified to serve on our board of directors due to his extensive experience in the

biotechnology industry and experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Stephen R. Biggar, M.D., Ph.D., has served as a member of our board of directors since October 2015. Since 2000, Dr. Biggar has been a partner at Baker Brothers. Dr. Biggar is currently Chairman of the board of directors of ACADIA Pharmaceuticals, serves on the board of Vivelix Pharmaceuticals, Ltd. and previously served on the board of directors of Synageva. Dr. Biggar holds an M.D. and a Ph.D. in Immunology from Stanford University and a BS in Genetics from the University of Rochester. We believe Dr. Biggar is qualified to serve on our board of directors due to his experience in the biotechnology industry, his medical and scientific training and experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Thomas R. Malley has served as a member of our board of directors since December 2016. Since May 2007, Mr. Malley has served as the President of Mossrock Capital, LLC, a private investment firm. Mr. Malley serves on the boards of directors of BeiGene, Ltd. and Kura Oncology, Inc., and previously served on the boards of directors of OvaScience, Inc., Cougar Biotechnology, Puma Biotechnology and Synageva. Mr. Malley holds a B.S. degree in Biology from Stanford University. We believe Mr. Malley is qualified to serve on our board of directors due to his experience working in the biopharmaceutical industry and experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Tracey L. McCain has served as a member of our board of directors since February 2018. Since September 2016, Ms. McCain has served as Executive Vice President and Chief Legal and Compliance Officer of Blueprint Medicine Corporation, or Blueprint, a biotechnology company. Prior to Blueprint, from January 2016 to September 2016, Ms. McCain was Senior Vice President and Head of Legal for Sanofi Genzyme, a global business unit of Sanofi,. Between joining Genzyme in May 1997 to January 2016, Ms. McCain held various roles at Genzyme, including as General Counsel following Genzyme's acquisition by Sanofi in 2011. Ms. McCain holds a J.D. from Columbia University School of Law and a B.A. from the University of Pennsylvania. We believe Ms. McCain is qualified to sit on our board of directors due to her experience working with numerous biotechnology and pharmaceutical companies.

Kimberly J. Popovits has served as a member of our board of directors since February 2018. Since 2009, Ms. Popovits has served as the Chief Executive Officer of Genomic Health, Inc., and since 2012, has served as the Chairman of the board of directors Ms. Popovits also serves on the board of directors of MyoKardia, Inc., and previously sat on the board of directors of ZS Pharma Inc. Ms. Popovits holds a B.A degree in Business from Michigan State University. We believe Ms. Popovits is qualified to sit on our board of directors due to her experience working with and serving on the boards of directors of numerous biotechnology and pharmaceutical companies.

Barry D. Quart, *Pharm.D.*, has served as a member of our board of directors since October 2015. Since 2013, Dr. Quart has served as the Chief Executive Officer and on the board of directors of Heron Therapeutics, Inc., or Heron, a biotechnology company. In 2006, Dr. Quart co-founded Ardea Biosciences, Inc., a biotechnology company, and served as its President and Chief Executive Officer, and on its board of directors, from its inception through May 2013. Dr. Quart previously served on the board of directors of Synageva. Dr. Quart holds a Pharm.D. degree from the University of California, San Francisco. We believe Dr. Quart is qualified to serve on our board of directors due to his extensive management experience in the biotechnology industry and his experience developing pharmaceutical products.

Family relationships

There are no family relationships between our board of directors and our executive officers.

Board composition

Our board of directors is currently comprised of seven members. Our directors hold office until the shareholders shall determine or, in the absence of such a determination, until the next annual general meeting or until their successors have been elected or appointed or their office is otherwise vacated.

Our amended and restated bye-laws provide that the authorized number of directors may be changed only by resolution of our board of directors. Our amended and restated bye-laws also provide that our directors may be removed only for cause by the affirmative vote of a majority of the votes entitled to be cast by our shareholders entitled to vote in an annual general election of directors. Any vacancy on our board of directors, including a vacancy resulting from an enlargement of our board of directors or from removal for cause not filled by the shareholders at the time, may be filled only by vote of a majority of our directors then in office.

In accordance with the terms of our amended and restated bye-laws, our board of directors is divided into three classes, Class I, Class II and Class III. At the first general meeting of shareholders following the initial public offering of our Class A common shares, each Class I director shall be elected for a three-year initial term of office, each Class II director shall be elected for a one-year initial term of office, and each Class III director shall be elected for a two-year initial term of office. Thereafter, members of each class shall serve three-year terms. The members of the classes are divided as follows:

- the Class I directors are Sanj K. Patel and Thomas R. Malley;
- the Class II directors are Stephen R. Biggar and Barry D. Quart; and
- the Class III directors are Felix J. Baker, Tracey L. McCain and Kimberly J. Popovits.

Upon the expiration of the term of a class of directors, directors in that class will be eligible to be elected for a new three-year term at the annual meeting of shareholders in the year in which their term expires.

We have no formal policy regarding board diversity. Our priority in selection of board members is identification of members who will further the interests of our shareholders through his or her established record of professional accomplishment, the ability to contribute positively to the collaborative culture among board members, knowledge of our business and understanding of the competitive landscape.

In selecting board members for nomination, our board may consider many factors, such as personal and professional integrity; experience in corporate management, such as serving as an officer or former officer of a pharmaceutical or biotechnology company; experience as a board member or executive officer of another publicly-held company; diversity of expertise and experience in substantive matters pertaining to our business relative to other board members; and diversity of background and perspective, including, but not limited to, with respect to age, gender, race, place of residence and specialized experience.

Director independence

Our board of directors has determined that, of our seven directors, Felix J. Baker, Stephen R. Biggar, Thomas R. Malley, Tracey L. McCain, Kimberly J. Popovits and Barry D. Quart do not have a relationship that would interfere with the exercise of independent judgment in carrying out the responsibilities of a director and that each of these directors is "independent" as that term is defined under the rules of The

Nasdaq Stock Market LLC. There are no family relationships among any of our directors or executive officers.

Board leadership structure

Our board of directors is currently chaired by our Chief Executive Officer, Sanj K. Patel, and our lead independent director is Felix J. Baker, Ph.D. Our corporate governance guidelines provide the flexibility for our board of directors to modify our leadership structure in the future as it deems appropriate.

Role of the board in risk oversight

One of the key functions of our board of directors is informed oversight of our risk management process. Our board of directors does not have a standing risk management committee, but rather administers this oversight function directly through our board of directors as a whole, as well as through various standing committees of our board of directors that address risks inherent in their respective areas of oversight. In particular, our board of directors is responsible for monitoring and assessing strategic risk exposure and our audit committee has the responsibility to consider and discuss our major financial risk exposures and the steps our management has taken to monitor and control these exposures, including guidelines and policies to govern the process by which risk assessment and management is undertaken. Our audit committee also monitors compliance with legal and regulatory requirements. Our nominating and corporate governance committee monitors the effectiveness of our corporate governance practices, including whether they are successful in preventing illegal or improper liability-creating conduct. Our compensation committee assesses and monitors whether any of our compensation policies and programs has the potential to encourage excessive risk-taking. While each committee is responsible for evaluating certain risks and overseeing the management of such risks, our entire board of directors is regularly informed through committee reports about such risks.

Board committees

Our board of directors has established three standing committees—audit, compensation and nominating and corporate governance—each of which operates under a charter that has been adopted by our board of directors. Each committee's charter is available under the Corporate Governance section of our website at *www.kiniksa.com*. The reference to our website address does not constitute incorporation by reference of the information contained at or available through our website, and you should not consider it to be a part of this prospectus.

Audit committee

The audit committee's responsibilities include:

- appointing, approving the compensation of, and assessing the independence of our registered public accounting firm;
- overseeing the work of our registered public accounting firm, including through the receipt and consideration of reports from such firm;
- reviewing and discussing with management and the registered public accounting firm our annual and quarterly financial statements and related disclosures;
- coordinating our board of directors' oversight of our internal control over financial reporting, disclosure controls and procedures and code of business conduct and ethics;

- discussing our risk management policies;
- meeting independently with our internal auditing staff, if any, registered public accounting firm and management;
- reviewing and approving or ratifying any related person transactions; and
- preparing the audit committee report required by SEC rules.

The members of our audit committee are Thomas R. Malley, Tracey L. McCain and Barry D. Quart. Mr. Malley serves as the chairperson of the committee. All members of our audit committee meet the requirements for financial literacy under the applicable listing rules of Nasdaq. Our board of directors has determined that Mr. Malley, Ms. McCain and Dr. Quart meet the independence requirements of Rule 10A-3 under the Exchange Act and the applicable Nasdaq rules. Our board of directors has determined that Mr. Malley is an "audit committee financial expert" as defined by applicable SEC rules and has the requisite financial sophistication as defined under the applicable Nasdaq rules.

Compensation committee

The compensation committee's responsibilities include:

- reviewing and approving, or recommending for approval by the board of directors, the compensation of our Chief Executive Officer and our other executive officers;
- overseeing and administering our cash and equity incentive plans;
- reviewing and making recommendations to our board of directors with respect to director compensation;
- reviewing and discussing annually with management our "Compensation Discussion and Analysis" to the extent required; and
- preparing the annual compensation committee report required by SEC rules, to the extent required.

The members of our compensation committee are Felix J. Baker, Kimberly J. Popovits and Barry D. Quart. Dr. Baker serves as the chairperson of the committee. Our board of directors has determined that Drs. Baker and Quart and Ms. Popovits are independent under the applicable Nasdaq rules, including the Nasdaq rules specific to membership on the compensation committee and that each is a "non-employee director" as defined in Rule 16b-3 promulgated under the Exchange Act.

Nominating and corporate governance committee

The nominating and corporate governance committee's responsibilities include:

- identifying individuals qualified to become board members;
- recommending to our board of directors the persons to be nominated for election as directors and to each board committee; and
- overseeing a periodic evaluation of our board of directors.

The members of our nominating and corporate governance committee are Felix J. Baker, Stephen R. Biggar and Thomas R. Malley. Dr. Biggar serves as the chairperson of the committee. Our board of directors has determined that Drs. Biggar and Baker and Mr. Malley are independent under the applicable Nasdaq rules.

Compensation committee interlocks and insider participation

No member of our compensation committee is or has been our current or former officer or employee. None of our executive officers served as a director or a member of a compensation committee (or other committee serving an equivalent function) of any other entity, one of whose executive officers served as a director or member of our compensation committee during the fiscal year ended December 31, 2017.

Code of ethics and code of conduct

We have adopted a written code of business conduct and ethics that applies to our directors, officers and employees, including our principal executive officer, principal financial officer, principal accounting officer or controller, or persons performing similar functions. Our code of business conduct and ethics is available under the Corporate Governance section of our website at *www.kiniksa.com*. In addition, we intend to post on our website all disclosures that are required by law or the Nasdaq rules concerning any amendments to, or waivers from, any provision of the code. The reference to our website address does not constitute incorporation by reference of the information contained at or available through our website, and you should not consider it to be a part of this prospectus.

Executive and director compensation

This section discusses the material components of the executive compensation program for our executive officers who are named in the "2018 summary compensation table" below. In 2018, our "named executive officers" and their positions were as follows:

- Sanj K. Patel, our Chief Executive Officer and Chairman of the Board of Directors;
- Stephen Mahoney, our President and Chief Operating Officer; and
- John F. Paolini, M.D., Ph.D., our Chief Medical Officer.

This discussion may contain forward-looking statements that are based on our current plans, considerations, expectations and determinations regarding future compensation programs. Actual compensation programs that we adopt following the completion of this offering may differ materially from the currently adopted programs summarized in this discussion.

2018 summary compensation table

The following table sets forth information concerning the compensation of our named executive officers for the years ended December 31, 2017 and 2018.

Name and principal position	Year	Salary ($)	Option awards ($)(1)	Non-equity incentive plan compensation ($)	All other compensation ($)(2)	Total ($)
Sanj K. Patel	2018	740,469	5,814,588	742,500	11,000	7,308,557
Chief Executive Officer and Chairman of the Board	2017	700,000	644,950	140,000	10,800	1,495,750
Stephen Mahoney	2018	434,970	1,980,564	291,892	11,000	2,718,426
President and Chief Operating Officer	2017	405,000	219,629	81,000	10,800	716,429
John F. Paolini, M.D., Ph.D	2018	409,626	1,875,468	207,900	11,000	2,503,994
Chief Medical Officer	2017	380,000	275,358	114,000	10,800	780,158

(1) Amounts reflect the full grant-date fair value of share options granted during the year computed in accordance with ASC Topic 718, rather than the amounts paid to or realized by the named individual. We provide information regarding the assumptions used to calculate the value of all option awards in Note 8 to our consolidated financial statements included in this prospectus.

(2) Amount shown represents 401(k) matching contributions. For additional information, refer to the discussion in the "Narrative disclosure to summary compensation table" below under the heading "—Other elements of compensation—Retirement plans."

Narrative disclosure to summary compensation table

The primary elements of compensation for our named executive officers are base salary, annual performance bonuses and long-term equity-based compensation awards. The named executive officers also generally participate in employee benefit plans and programs that we offer to our other full-time employees on the same basis.

2018 salaries

We pay our named executive officers a base salary that is intended to provide a fixed component of compensation reflecting the executive's skill set, experience, role and responsibilities. Base salaries for our named executive officers generally have been set at levels deemed necessary to attract and retain the

named executive officers. Our board of directors approved increases in the base salaries of our named executive officers effective May 30, 2018. The following table shows the annual base salaries of our named executive officers for 2018 before and after the May 30, 2018 increases:

Name	2018 Annual Base Salary Before Salary Increase ($)	2018 Annual Base Salary After Salary Increase ($)
Sanj K. Patel	725,000	750,000
Stephen Mahoney	421,200	442,260
John F. Paolini, M.D., Ph.D.	391,400	420,000

In December 2018, our compensation committee approved the following increased annual base salaries for our named executive officers, effective January 1, 2019: $780,000 for Mr. Patel, $477,641 for Mr. Mahoney and $443,100 for Dr. Paolini.

2018 bonuses

We offer our named executive officers the opportunity to earn annual performance bonuses to compensate them for attaining short-term company and individual goals as approved by our board of directors. For 2018, performance bonuses were based on attaining corporate goals relating to the overall business, generally comprised of goals with respect to: (i) the advancement and growth of our portfolio of products (carrying an 80% weighting), including development of product candidates, business development, intellectual property protection, manufacturing and supply, and organizational and corporate structure; (ii) capitalization, financial management, and capital allocation (carrying a 15% weighting); and (iii) compliance and training (carrying a 5% weighting). The 2018 target bonus amounts for our named executive officers, expressed as percentages of their respective annual base salaries, were 60% for Mr. Patel, 40% for Mr. Mahoney and 30% for Dr. Paolini.

In December 2018, our compensation committee reviewed performance against the 2018 bonus goals and approved cash bonuses for the named executive officers in the amounts set forth in the Non-Equity Incentive Plan Compensation column of the 2018 Summary Compensation Table above. In December 2018, our compensation committee also approved the following 2019 target bonus amounts for our named executive officers, expressed as percentages of their respective annual base salaries: 65% for Mr. Patel, 45% for Mr. Mahoney and 35% for Dr. Paolini.

Equity compensation

We generally offer share options to our employees, including our named executive officers, as the long-term incentive component of our compensation program. Share options allow our employees to purchase our Class A common shares at a price equal to the fair market value per Class A common share on the date of grant, as determined by the board of directors. In 2018, our named executive officers were granted the share options set forth in the table below. These share options were granted with exercise prices equal to the fair market value of our Class A common shares on the date of grant. Refer to our

Outstanding equity awards at 2018 fiscal year-end table for additional information on the share option grants made in 2018.

Named executive officer	2018 option awards granted
Sanj K. Patel	564,182
Stephen Mahoney	178,094
John F. Paolini, M.D., Ph.D.	173,094

Prior to our IPO, we issued share options under our 2015 Equity Incentive Plan, or the 2015 Plan.

Effective on the effective date of the registration statement for our IPO, our board of directors adopted, and our shareholders approved, the 2018 Incentive Award Plan, or the 2018 Plan, in order to facilitate the grant of cash and equity incentives to directors, employees (including our named executive officers) and consultants and to enable us to obtain and retain services of these individuals, which we believe is essential to our long-term success. Following effectiveness of the 2018 Plan, we ceased making grants under our 2015 Plan. However, the 2015 Plan continues to govern the terms and conditions of the outstanding awards granted under it. For additional information about the 2015 Plan and the 2018 Plan, please see the section titled "Executive compensation plans" below.

Other elements of compensation

Retirement plans

We maintain a 401(k) retirement savings plan for our employees, including our named executive officers, who satisfy certain eligibility requirements. Our named executive officers are eligible to participate in the 401(k) plan on the same terms as other full-time employees. We provide matching contributions of 100% of the first 3% of each participant's salary contributed, plus 50% for each of the next 2% contributed. Employee contributions are allocated to each participant's individual account and are then invested in selected investment alternatives according to the participants' directions. Employees are immediately and fully vested in their own contributions and the employer match. We believe that providing a vehicle for tax-deferred retirement savings though our 401(k) plan adds to the overall desirability of our executive compensation package and further incentivizes our employees, including our named executive officers, in accordance with our compensation policies.

Employee benefits and perquisites

All of our full-time employees, including our named executive officers, are eligible to participate in our health and welfare plans, including medical, dental and vision benefits, a healthcare spending account, a dependent care flexible spending account, short-term and long-term disability insurance and life insurance to the same extent as our other full-time employees generally, subject to the terms and eligibility requirements of those plans.

No tax gross-ups

We do not make gross-up payments to cover our named executive officers' personal income taxes that may pertain to any of the compensation or perquisites paid or provided by us.

Outstanding equity awards at 2018 fiscal year-end

The following table summarizes the number of Class A common shares underlying outstanding equity incentive plan awards for each named executive officer as of December 31, 2018.

		Option awards(1)				Share awards	
Name	Vesting start date	Number of securities underlying unexercised options (#) exercisable	Number of securities underlying unexercised options (#) unexercisable	Option exercise price ($)	Option expiration date	Number of shares that have not vested (#)	Value of shares that have not vested ($)
Sanj K. Patel	8/1/2015	419,809	83,959(2)	1.59	12/15/2025	—	—
	6/28/2017	96,740	161,229(2)	3.80	6/28/2027	—	—
	3/1/2018	—	439,182(3)	10.36	3/1/2028	—	—
	9/20/2018	—	125,000(2)	30.93	9/20/2028	—	—
	—	—	—	—	—	36,600(4)	1,028,094
	—	—	—	—	—	182,994(5)	5,140,301
Stephen Mahoney	8/1/2015	104,952	20,989(2)	1.59	12/15/2025	—	—
	6/28/2017	32,944	54,904(2)	3.80	6/28/2027	—	—
	3/1/2018	—	128,094(3)	10.36	3/1/2028	—	—
	9/20/2018	—	50,000(2)	30.93	9/20/2028	—	—
	—	—	—	—	—	18,300(4)	514,047
	—	—	—	—	—	91,497(5)	2,570,151
John F. Paolini, M.D., Ph.D	8/15/2016	132,754	94,820(2)	1.86	9/13/2026	—	—
	6/28/2017	41,303	68,835(2)	3.80	6/28/2027	—	—
	3/1/2018	—	128,094(3)	10.36	3/1/2028	—	—
	9/20/2018	—	45,000(2)	30.93	9/20/2028	—	—

(1) Pursuant to each named executive officer's employment agreement, in the event of a termination of employment by the Company without Cause or a result of the named executive officer's death, disability or resignation for Good Reason (as such capitalized terms are defined in their respective employment agreements), Mr. Mahoney and Dr. Paolini are entitled to accelerated vesting of all of their then-unvested Company equity or equity-based awards that would have, absent termination, become vested within 12 months following termination, and Mr. Patel is entitled to accelerated vesting of all of his then-unvested equity or equity-based awards that would have, absent termination, become vested within 18 months following termination. In addition, in the event of a change in control (as defined in the applicable option award agreement), each named executive officer will become immediately 100% fully vested in the named executive officer's option to the extent that such award is not assumed or substituted.

(2) The options vest over a four-year period with 25% of the shares vesting on the first anniversary of the corresponding vesting start date, and 2.0833% of the shares vesting monthly for three years thereafter.

(3) The options vest over a six-year period with 16% of the shares vesting on the first anniversary of the corresponding vesting start date, 48% of the shares vesting in 36 equal monthly installments over the following three years and 36% of the shares vesting in 24 equal monthly installments over the two years thereafter.

(4) Represents unvested Class A common shares that were acquired by the named executive officer under a restricted stock agreement in August 2015. The unvested shares will vest in equal monthly installments through August 1, 2019, subject to accelerated vesting if, following a Sale, the named executive officer's employment is terminated by us without Cause or by the named executive officer for Good Reason (as such capitalized terms are defined in the applicable restricted stock agreement). The number of unvested Class A common shares shown for Mr. Patel includes 18,300 unvested shares that have been transferred by Mr. Patel to the Manisha S. Patel 2016 Irrevocable Trust. The number of unvested Class A common shares shown for Mr. Mahoney includes 6,099 unvested shares that have been transferred by Mr. Mahoney to the Stephen F. Mahoney 2016 Irrevocable Trust.

(5) Represents unvested Class B common shares issued to the named executive officer in respect of, and subject to the same vesting and other conditions as, restricted Class A common shares acquired by the named executive officer under a restricted stock agreement in August 2015. The unvested shares will vest in equal monthly installments through August 1, 2019, subject to accelerated vesting if, following a Sale, the named executive officer's employment is terminated by us without Cause or by the named executive officer for Good Reason (as such capitalized terms are defined in the applicable restricted stock agreement).

Executive compensation arrangements

We have entered into employment agreements with each of our named executive officers. Certain key terms of these agreements are described below.

Sanj K. Patel

The term of our employment agreement with Mr. Patel lasts until either the Company or Mr. Patel terminates his employment by giving notice to the other party or his employment terminates due to his death. Pursuant to the employment agreement, Mr. Patel is entitled to receive an annual base salary of at least $750,000, subject to increase from time to time by the Company and the opportunity to earn an annual performance-based bonus based on actual corporate and individual performance against established objectives for each calendar year, with a minimum target bonus opportunity of 60% of his annual base salary. In addition, if Mr. Patel's employment with us is terminated as a result of his death or disability, by the Company without Cause, or by Mr. Patel for Good Reason, whether or not in connection with a change in control, he will be entitled to receive (a) a lump sum payment equal to (i) 200% of the sum of his annual base salary and the target bonus for the year of termination plus (ii) $25,000 and (b) a prorated portion of his target bonus for the year of termination. Also, if the termination occurs other than during the 12-month period following a change in control, Mr. Patel will be entitled to accelerated vesting of all of his then-unvested time-vesting equity that would have, absent termination, become vested within 18 months following termination, or if the termination occurs during the 12-month period following a change in control, Mr. Patel will be entitled to full accelerated vesting of all of his then-unvested time-vesting equity. Mr. Patel's right to receive these severance payments and benefits is subject to his execution and non-revocation of a release of claims for the benefit of the Company and his compliance with certain confidentiality obligations and restrictive covenants.

In the event of a change in control, Mr. Patel will become immediately 100% fully vested in each time-vesting equity award granted to him that is not assumed or substituted for in the change in control transaction.

Stephen Mahoney

The term of our employment agreement with Mr. Mahoney lasts until either the Company or Mr. Mahoney terminates his employment by giving notice to the other party or his employment terminates due to his death. Pursuant to the employment agreement, Mr. Mahoney is entitled to receive an annual base salary of $442,260, subject to change from time to time by the Company, and the opportunity to earn an annual performance-based bonus, with an initial target bonus opportunity of 40% of his annual base salary. In addition, if Mr. Mahoney's employment with us is terminated as a result of his death or disability, by the Company without Cause, or by Mr. Mahoney for Good Reason, he will be entitled to receive (a) a lump sum payment equal to his annual base salary plus $16,500, (b) a prorated portion (or, if the termination occurs during the 12 months following a change in control, 100%) of his target bonus for the year of termination and (c) accelerated vesting of all of his then-unvested time-vesting equity that would have, absent termination, become vested within 12 months following termination (or, if the termination occurs during the 12 months following a change in control, full accelerated vesting of all of his then-unvested time-vesting equity). Mr. Mahoney's right to receive these severance payments and benefits is subject to his execution and non-revocation of a release of claims for the benefit of the Company and his compliance with certain confidentiality obligations and restrictive covenants.

John F. Paolini, M.D., Ph.D.

The term of our employment agreement with Dr. Paolini lasts until either the Company or Dr. Paolini terminates his employment by giving notice to the other party or his employment terminates due to his death. Pursuant to the employment agreement, Dr. Paolini is entitled to receive an annual base salary of

$420,000, subject to change from time to time by the Company, and the opportunity to earn a discretionary performance-based bonus, with a target bonus opportunity of 30% of his annual base salary. In addition, if Dr. Paolini's employment with us is terminated as a result of his death or disability or without Cause, he will be entitled to receive (a) a lump sum payment that is equivalent to 9 months of his annual base salary (or, if the termination occurs during the 12 months following a change in control, a lump sum payment that is equivalent to 12 months of his annual base salary) plus $16,500, (b) a prorated portion (or, if the termination occurs during the 12 months following a change in control, 100%) of his target bonus for the year of termination and (c) accelerated vesting of all of his then-unvested time-vesting equity that would have, absent termination, become vested within 12 months following termination (or, if the termination occurs during the 12 months following a change in control, full accelerated vesting of all of his then-unvested time-vesting equity). Dr. Paolini's right to receive these severance payments and benefits is subject to his execution and non-revocation of a release of claims for the benefit of the Company and his compliance with certain confidentiality obligations and restrictive covenants.

As used in the executive employment agreements, the following capitalized terms generally have the following meanings:

- The term Cause generally means (i) gross negligence or willful misconduct in performance of the named executive officer's duties which results in material damage to us; (ii) the commission of any act of fraud, embezzlement or professional dishonesty with respect to our business; (iii) the commission of a felony or crime involving moral turpitude; (iv) the material breach of any provision of the executive employment agreement or any other written agreement between the named executive officer and us; or (v) the failure to comply with our lawful directives, which results in damage to us.

- The term Good Reason generally means the occurrence of any of the following events without the named executive officer's written consent: (i) a demotion or, in the case of Mr. Patel only, the assignment of duties materially inconsistent with his title, position, status, reporting relationships, authority, duties or responsibilities with us; (ii) a requirement that the named executive officer relocate his primary reporting location to a location more than fifty (50) miles from our offices in Lexington, Massachusetts; (iii) our breach of the executive employment agreement with the named executive officer; (iv) our failure to comply with the provisions addressing each named executive officer's compensation and benefits, including the base salary, bonus compensation, and annual vacation, other than insubstantial or inadvertent failures not in bad faith that we remedy promptly after receiving notice thereof; (v) for Mr. Patel only, a material diminution in the budget over which he has responsibility; or (vi) for Mr. Mahoney only, a reduction of more than five percent of his base salary other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated.

Director compensation

Directors who are also our employees do not receive compensation for their service as directors. Prior to our IPO, directors who were affiliated with one of our principal shareholders did not receive compensation for their service as directors. For the portion of 2018 preceding our IPO, our other non-employee directors received cash payments of $10,000 per year, paid quarterly, and awards of our share options as compensation for their service as directors.

Effective on the effective date of the registration statement for our IPO, we adopted a compensation program for our non-employee directors under which each non-employee director receives the following amounts for their services on our board of directors:

- an option to purchase 37,965 Class A common shares upon the director's initial election or appointment to our board of directors that occurs after our initial public offering,

- if the director has served on our board of directors for at least six months as of the date of an annual meeting of shareholders, an option to purchase 18,760 Class A common shares on the date of the annual meeting,

- an annual director fee of $35,000, and

- if the director serves on a committee of our board of directors or in the other capacities stated below, an additional annual fee as follows:

- chairman of the board or lead independent director, $22,500,
- chairman of the audit committee, $15,000,
- audit committee member other than the chairman, $7,500,
- chairman of the compensation committee, $10,000,
- compensation committee member other than the chairman, $5,000,
- chairman of the nominating and corporate governance committee, $8,000, and
- nominating and corporate governance committee member other than the chairman, $4,000.

Share options granted to our non-employee directors under the program have an exercise price equal to the fair market value of our Class A common shares on the date of grant and expire not later than ten years after the date of grant. The share options granted upon a director's initial election or appointment vest and become exercisable as to one-third of the shares on the first anniversary of the date of grant and as to the remainder in twenty-four substantially equal monthly installments thereafter, subject to the director continuing in service through each such vesting date. The share options granted annually to directors vest and become effective in twelve substantially equal monthly installments following the date of grant, subject to the director continuing in service through each such vesting date. In addition, all unvested share options will vest in full upon the occurrence of a change in control.

Director fees under the program are payable in arrears in four equal quarterly installments not later than the fifteenth day following the final day of each calendar quarter, provided that the amount of each payment are prorated for any portion of a quarter that a director is not serving on our board and no fee is payable in respect of any period prior to the effective date of the registration statement for our initial public offering of Class A common shares.

2018 director compensation table

The following table sets forth in summary form information concerning the compensation that was earned by or paid to each of our non-employee directors during the year ended December 31, 2018:

Name	Fees earned or paid in cash ($)	Option awards ($)(1)	Total ($)
Felix J. Baker, Ph.D	41,708	216,640	258,348
Stephen R. Biggar, M.D., Ph.D	25,083	216,640	241,723
Thomas R. Malley	35,667	134,476	170,143
Tracey L. McCain	28,958	317,752	346,710
Kimberly J. Popovits	27,500	317,752	345,252
Barry D. Quart, Pharm.D	31,875	134,476	166,351

(1) Amounts reflect the full grant-date fair value of share options granted during 2018 computed in accordance with ASC Topic 718, rather than the amounts paid to or realized by the named individual. We provide information regarding the assumptions used to calculate the value of all option awards in Note 8 to our consolidated financial statements included in this prospectus.

The following table sets forth the aggregate numbers of share options (exercisable and unexercisable) held by our non-employee directors as of December 31, 2018. Refer to our Outstanding equity awards at 2018 fiscal year-end table for information regarding equity awards held by Mr. Patel as of December 31, 2018.

Name	Option awards(#)
Felix J. Baker, Ph.D	18,760
Stephen R. Biggar, M.D., Ph.D	18,760
Thomas R. Malley	69,536
Tracey L. McCain	45,748
Kimberly J. Popovits	45,748
Barry D. Quart, Pharm.D	69,536

For the portion of 2018 preceding our IPO, our non-employee directors who were not affiliated with one of our principal stockholders received awards of share options as a component of their compensation for their service as directors. In March 2018, Mr. Malley and Dr. Quart received grants of options to purchase 20,129 of our Class A common shares and Mses. McCain and Popovits received grants of options to purchase 45,748 of our Class A common shares. These shares options have exercise prices equal to $10.36, which the board of directors determined was the fair market value per Class A common share on the date of grant. The share options granted to Mr. Malley and Dr. Quart vest in twelve equal monthly installments following the effective date of grant. The share options granted to Mses. McCain and Popovits vest as to one-third of the underlying shares on March 1, 2019 and as to the remaining shares in 24 equal monthly installments thereafter. In May 2018, Drs. Baker and Biggar received grants of options to purchase 18,760 of our Class A common shares. These share options have exercise prices equal to the initial public offering price of $18.00 per Class A common share in our IPO and vest in twelve equal monthly installments following the effective date of grant.

Executive compensation plans

The following summarizes the material terms of the long-term incentive compensation plans in which our named executive officers and directors are eligible to participate, and the 2015 Plan, under which we have

previously made periodic grants of equity and equity-based awards to our named executive officers and directors.

2015 equity incentive plan

Until May 23, 2018 (the effective date of the 2018 Plan), the 2015 Plan provided for our company to grant qualified incentive options, nonqualified options, share grants and other share-based awards to employees, directors, and consultants to purchase our company's Class A common shares. On May 23, 2018, our company ceased granting awards under the 2015 Plan. At that time, the 4,691,213 shares of Class A common shares subject to outstanding awards under the 2015 Plan remained reserved for issuance under the plan pursuant such awards and the 92,170 shares of Class A common shares that had been available for future grant under the 2015 Plan were no longer authorized and reserved for issuance or available for future grant under the 2015 Plan. However, the 2015 Plan will continue to govern the terms and conditions of the outstanding awards granted under it. Our Class A common shares subject to awards granted under the 2015 Plan that are forfeited, lapse unexercised or are settled in cash will be available for issuance under the 2018 Plan.

Administration. Our board of directors administers the 2015 Plan and has the authority to interpret the provisions of the 2015 Plan and awards outstanding thereunder, to make all rules and determinations which it deems necessary or advisable for the administration of the 2015 Plan, and to make all other determinations in the judgment of the board of directors that are necessary and desirable for the administration of the 2015 Plan. Prior to May 23, 2018, our board of directors had the authority to determine which employees, directors and consultants would be granted awards, to determine the number of shares for which awards would be granted, to specify the terms and conditions upon which awards may be granted, to specify the terms and conditions of award agreements under the 2015 Plan, and to make all other determinations in the judgment of the board of directors that are necessary and desirable for the administration of the 2015 Plan. The board of directors has delegated its general administrative authority under the 2015 Plan to its compensation committee.

Types of awards. The 2015 Plan provided for the grant of share options, including share options intended to qualify as incentive stock options, or ISOs, under the U.S. Internal Revenue Code of 1986, as amended, or the Code, share grants and share-based awards to employees, directors and consultants of the company or its affiliates, except that share options intended to qualify as ISOs may only have been granted to employees who are residents of the United States.

Certain transactions. If certain changes are made in, or events occur with respect to, our Class A common shares, the 2015 Plan and outstanding awards will be adjusted in the class, number and, as applicable, exercise price of securities as determined by the board of directors. In the event of certain corporate transactions of our company, including an amalgamation, consolidation, merger or sale of all or substantially all of our assets, our board or the board of directors of any corporation assuming the obligations under the 2015 Plan, may, in its discretion, take any one or more of the following actions, as to some or all options or share-based awards outstanding under the 2015 Plan (and need not take the same action as to each such option or share-based award): (i) make appropriate provisions for the continuation of options by substituting on an equitable basis for the shares then subject to options either the consideration payable with respect to the outstanding Class A common shares or securities of any successor or acquiring entity; (ii) upon written notice to the participants, provide that the options will terminate unless they are exercised within a specified number of days of the date of such notice; (iii) terminate the options in exchange for payment of an amount equal to the consideration payable upon consummation of such transaction to a holder of the number of Class A common shares into which such

option would have been exercisable, less the aggregate exercise price; (iv) make appropriate provision for the continuation of such share grants on the same terms and conditions by substituting on an equitable basis for the shares then subject to the share grants either the consideration payable with respect to such outstanding shares in connection with the transaction or securities of any successor or acquiring entity; and (v) provide that, upon consummation of the transaction, each outstanding share grant shall be terminated in exchange for a payment of an amount equal to the consideration payable upon consummation of such transaction to a holder of the number of Class A common shares comprising the share grant.

Amendment and termination. The board of directors may terminate, modify or amend the 2015 Plan from time to time, provided that any amendment or modification may not adversely affect the rights of a holder of an outstanding award without such holder's consent. The board of directors may amend or modify the 2015 Plan and any outstanding ISOs to the extent necessary to qualify any or all such options for favorable federal income tax treatment; however, any amendment approved by the board of directors which is determined to be of a scope that requires shareholder approval will be subject to obtaining such approval before taking effect.

2018 incentive award plan

Our board of directors and our shareholders have approved the 2018 Plan, under which we may grant cash and equity-based incentive awards to eligible service providers in order to attract, retain and motivate the persons who make important contributions to our company. The material terms of the 2018 Plan are summarized below.

Eligibility and administration

Our employees, consultants and directors, and employees and consultants of our subsidiaries, are eligible to receive awards under the 2018 Plan. The 2018 Plan is administered by our board of directors, which has delegated its duties and responsibilities to one or more committees of our directors and/or officers (referred to collectively as the plan administrator below), subject to the limitations imposed under the 2018 Plan, Section 16 of the Exchange Act, stock exchange rules and other applicable laws. The plan administrator has the authority to take all actions and make all determinations under the 2018 Plan, to interpret the 2018 Plan and award agreements and to adopt, amend and repeal rules for the administration of the 2018 Plan as it deems advisable. The plan administrator also has the authority to grant awards, determine which eligible service providers receive awards, and set the terms and conditions of all awards under the 2018 Plan, including any vesting and vesting acceleration provisions, subject to the conditions and limitations in the 2018 Plan. Our board of directors has delegated its general administrative authority under the 2018 Plan to its compensation committee.

Shares available for awards

An aggregate of 4,466,500 Class A common shares were initially available for issuance under the 2018 Plan. The number of Class A common shares initially available for issuance will be increased by an annual increase on January 1 of each calendar year beginning in 2019 and ending in and including 2028, equal to the lesser of (A) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (B) a smaller number of our Class A common shares determined by our board of directors. No more than 27,915,000 Class A common shares may be issued under the 2018 Plan upon the exercise of ISOs. Shares issued under the 2018 Plan may be designated but unissued shares, shares purchased on the open market or treasury shares.

If an award under the 2018 Plan or the 2015 Plan, expires, lapses or is terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited, any unused shares subject to the award will, as applicable, become or again be available for new grants under the 2018 Plan. Awards granted under the 2018 Plan in substitution for any options or other share or share-based awards granted by an entity before the entity's merger or consolidation with us or our acquisition of the entity's property or shares will not reduce the shares available for grant under the 2018 Plan, but may count against the maximum number of shares that may be issued upon the exercise of ISOs.

As of December 31, 2018, options to purchase 5,960,939 Class A common shares were outstanding and 3,175,665 shares were available for future issuance under the 2018 Plan.

Awards

The 2018 Plan provides for the grant of options to purchase shares, including ISOs, non-qualified share options, or NSOs, share appreciation rights, or SARs, restricted shares, dividend equivalents, restricted share units, or RSUs, and other share or cash based awards. Certain awards under the 2018 Plan may constitute or provide for payment of "nonqualified deferred compensation" under Section 409A of the Code. All awards under the 2018 Plan will be set forth in award agreements, which will detail the terms and conditions of awards, including any applicable vesting and payment terms and post-termination exercise limitations. A brief description of each award type follows.

- *Share options and SARs.* Share options provide for the purchase of our Class A common shares in the future at an exercise price set on the grant date. ISOs, by contrast to NSOs, may provide tax deferral beyond exercise and favorable capital gains tax treatment to their holders if certain holding period and other requirements of the Code are satisfied. SARs entitle their holder, upon exercise, to receive from us an amount equal to the appreciation of the shares subject to the award between the grant date and the exercise date. The plan administrator determines the number of shares covered by each option and SAR, the exercise price of each option and SAR and the conditions and limitations applicable to the exercise of each option and SAR. The exercise price of a share option or SAR will not be less than 100% of the fair market value of the underlying share on the grant date (or 110% in the case of ISOs granted to certain significant shareholders), except with respect to certain substitute awards granted in connection with a corporate transaction. The term of a share option or SAR may not be longer than ten years (or five years in the case of ISOs granted to certain significant shareholders).

- *Restricted shares and RSUs.* Restricted shares are an award of nontransferable Class A common shares that remain forfeitable unless and until specified conditions are met and which may be subject to a purchase price. RSUs are contractual promises to deliver Class A common shares in the future, which may also remain forfeitable unless and until specified conditions are met and may be accompanied by the right to receive the equivalent value of dividends paid our Class A common shares prior to the delivery of the underlying shares. The plan administrator may provide that the delivery of the shares underlying RSUs will be deferred on a mandatory basis or at the election of the participant. The terms and conditions applicable to restricted shares and RSUs are determined by the plan administrator, subject to the conditions and limitations contained in the 2018 Plan.

- *Other share or cash based awards.* Other share or cash based awards are awards of cash, fully vested Class A common shares and other awards valued wholly or partially by referring to, or otherwise based on, our Class A common shares or other property. Other share or cash based awards may be granted to participants and may also be available as a payment form in the settlement of other awards, as standalone payments and as payment in lieu of compensation to which a participant is otherwise entitled. The plan administrator determines the terms and conditions of other share or cash based

awards, which may include any purchase price, performance goal, transfer restrictions and vesting conditions.

Certain transactions

In connection with certain corporate transactions and events affecting our Class A common shares, including a change in control, or change in any applicable laws or accounting principles, the plan administrator has broad discretion to take action under the 2018 Plan to prevent the dilution or enlargement of intended benefits, facilitate the transaction or event or give effect to the change in applicable laws or accounting principles. This includes canceling awards for cash or property, accelerating the vesting of awards, providing for the assumption or substitution of awards by a successor entity, adjusting the number and type of shares subject to outstanding awards and/or with respect to which awards may be granted under the 2018 Plan and replacing or terminating awards under the 2018 Plan. In addition, in the event of certain non-reciprocal transactions with our shareholders, the plan administrator will make equitable adjustments to the 2018 Plan and outstanding awards as it deems appropriate to reflect the transaction.

Plan amendment and termination

Our board of directors may amend or terminate the 2018 Plan at any time; however, no amendment, other than an amendment that increases the number of shares available under the 2018 Plan, may materially and adversely affect an award outstanding under the 2018 Plan without the consent of the affected participant and shareholder approval will be obtained for any amendment to the extent necessary to comply with applicable laws. Further, the plan administrator cannot, without the approval of our shareholders, amend any outstanding share option or SAR to reduce its price per share other than in the context of corporate transactions or equity restructurings, as described above. The 2018 Plan will remain in effect until the tenth anniversary of the earlier of the date that our board of directors or our shareholders approve the 2018 Plan, unless earlier terminated by our board of directors. No awards may be granted under the 2018 Plan after its termination.

Foreign participants, claw-back provisions, transferability and participant payments

The plan administrator may modify awards granted to participants who are foreign nationals or employed outside the United States or establish subplans or procedures to address differences in laws, rules, regulations or customs of such foreign jurisdictions. All awards will be subject to any company claw-back policy as set forth in such claw-back policy, the 2018 Plan or the applicable award agreement. Except as the plan administrator may determine or provide in an award agreement, awards under the 2018 Plan are generally non-transferrable, except by will or the laws of descent and distribution, or, subject to the plan administrator's consent, pursuant to a domestic relations order, and are generally exercisable only by the participant. With regard to tax withholding obligations arising in connection with awards under the 2018 Plan and exercise price obligations arising in connection with the exercise of share options under the 2018 Plan, the plan administrator may, in its discretion, accept cash, wire transfer or check, our Class A common shares that meet specified conditions, a promissory note, a "market sell order," such other consideration as the plan administrator deems suitable or any combination of the foregoing.

2018 employee share purchase plan

Our board of directors and our shareholders have approved the 2018 Employee Share Purchase Plan, or the 2018 ESPP. The material terms of the 2018 ESPP are summarized below.

Shares available for awards; administration

A total of 670,000 Class A common shares were initially available for issuance under the 2018 ESPP. In addition, the number of Class A common shares available for issuance under the 2018 ESPP will be annually increased on January 1 of each calendar year beginning in 2019 and ending in and including 2028, by an amount equal to the lesser of (A) 1% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (B) such smaller number of Class A common shares as is determined by our board of directors, provided that no more than 6,420,000 Class A common shares may be issued under the 2018 ESPP. Our board of directors or a committee of our board of directors administers and has authority to interpret the terms of a 2018 ESPP and determine eligibility of participants. Our board of directors has delegated its general administrative authority under the 2018 ESPP to its compensation committee.

As of December 31, 2018, 648,660 Class A common shares were available for future issuance under the 2018 ESPP. In December 2018, our board of directors determined that the January 1, 2019 automatic increase in shares available under the 2018 ESPP would be zero shares.

Eligibility

Our employees are eligible to participate in the 2018 ESPP if they are customarily employed by us or a participating subsidiary for more than twenty hours per week and more than five months in any calendar year. However, an employee may not be granted rights to purchase shares under our 2018 ESPP if the employee, immediately after the grant, would own (directly or through attribution) shares possessing 5% or more of the total combined voting power or value of all classes of our shares.

Grant of rights

The 2018 ESPP is intended to qualify under Section 423 of the Code and shares will be offered under the 2018 ESPP during offering periods. The length of the offering periods under the 2018 ESPP are determined by the plan administrator and may be up to twenty-seven months long. Employee payroll deductions will be used to purchase shares on each purchase date during an offering period. The purchase dates for each offering period will be the final trading day in the offering period. Offering periods under the 2018 ESPP will commence when determined by the plan administrator. The plan administrator may, in its discretion, modify the terms of future offering periods.

The 2018 ESPP permits participants to purchase our Class A common shares through payroll deductions of up to a specified percentage of their eligible compensation. The plan administrator establishes a maximum number of shares that may be purchased by a participant during any offering period, which, in the absence of a contrary designation, is 25,000 shares. In addition, no employee may accrue the right to purchase shares under the 2018 ESPP at a rate in excess of $25,000 worth of shares during any calendar year during which such a purchase right is outstanding (based on the fair market value per share of our Class A common shares as of the first day of the offering period).

On the first trading day of each offering period, each participant is automatically granted an option to purchase our Class A common shares. The option expires at the end of the applicable offering period, and is exercised at that time to the extent of the payroll deductions accumulated during the offering period. The purchase price of the shares, in the absence of a contrary designation, is 85% of the lower of the fair market value of our Class A common shares on the first trading day of the offering period or on the purchase date. Participants may voluntarily end their participation in the 2018 ESPP at any time not later than a specified period prior to the end of the applicable offering period, and are paid their accrued

payroll deductions that have not yet been used to purchase shares of our Class A common shares. Participation ends automatically upon a participant's termination of employment.

A participant may not transfer rights granted under the 2018 ESPP other than by will or the laws of descent and distribution.

Certain transactions

In the event of certain non-reciprocal transactions or events affecting our Class A common shares, the plan administrator may make equitable adjustments to the 2018 ESPP and outstanding rights. In the event of certain unusual or non-recurring events or transactions, including a change in control, the plan administrator may provide for (1) either the replacement of outstanding rights with other rights or property or termination of outstanding rights in exchange for cash, (2) the assumption or substitution of outstanding rights by the successor or survivor corporation or parent or subsidiary thereof, if any, (3) the adjustment in the number and type of shares subject to outstanding rights, (4) the use of participants' accumulated payroll deductions to purchase shares on a new purchase date prior to the next scheduled purchase date and termination of any rights under ongoing offering periods or (5) the termination of all outstanding rights.

Plan amendment

The plan administrator may amend, suspend or terminate the 2018 ESPP at any time. However, shareholder approval will be obtained for any amendment that increases the aggregate number or changes the type of shares that may be sold pursuant to rights under the 2018 ESPP, changes the corporations or classes of corporations whose employees are eligible to participate in the 2018 ESPP or changes the 2018 ESPP in any manner that would cause the 2018 ESPP to no longer be an employee share purchase plan within the meaning of Section 423(b) of the Code.

Certain relationships and related person transactions

The following includes a summary of transactions since our inception in July 2015 to which we have been a party in which the amount involved exceeded or will exceed $120,000, and in which any of our directors, executive officers or, to our knowledge, beneficial owners of more than 5% of any class of our voting shares or any member of the immediate family of any of the foregoing persons had or will have a direct or indirect material interest, other than equity and other compensation, termination, change in control and other arrangements, which are described under "Executive and director compensation." We also describe below certain other transactions with our directors, executive officers and shareholders.

Concurrent private placement

One or more entities managed by Baker Brothers have agreed to purchase 2,000,000 of our non-voting Class A1 common shares in a private placement exempt from the registration requirements of the Securities Act at a sale price equal to the public offering price of our Class A common shares in this offering. The consummation of the concurrent private placement is contingent on the closing of this offering and the satisfaction of certain other customary conditions. The underwriters will serve as placement agents for the concurrent private placement and receive a placement agent fee equal to a percentage of the total purchase price of the private placement shares, which percentage will be equal to the percentage discount the underwriters will receive on shares sold in this public offering. The concurrent private placement is expected to close concurrently with this offering. The consummation of this offering is not contingent on the consummation of the concurrent private placement.

Participation in our initial public offering

Baker Brothers and HH RSV-XVII Holdings Limited, each a beneficial owner of more than 5% of our Class A common shares, purchased 3,000,000 and 1,388,888 Class A common shares, respectively, in our initial public offering at the initial public offering price of $18.00 per share.

Preferred share financings

Series A preferred share financing

In October 2015, we issued and sold an aggregate of 8,028,809 Series A Preferred Shares, and in September 2016 we sold an additional 9,099,311 Series A Preferred Shares to new investors and certain of our directors and executive officers at a price of $4.6707 per share, resulting in aggregate gross proceeds of $80.0 million.

Series B preferred share financing

In March 2017, we issued and sold an aggregate of 5,757,372 Series B Preferred Shares to new investors, existing shareholders, a director and an executive officer at a price of $6.9475 per share, resulting in aggregate gross proceeds of $40.0 million.

Series C preferred share financing

In February 2018, we issued and sold an aggregate of 12,784,601 Series C Preferred Shares to new investors, existing shareholders and certain executive officers at a price of $15.6438 per share, resulting in aggregate gross proceeds of $200.0 million.

The following table sets forth the aggregate number of preferred shares acquired by the listed holders of more than 5% of any class of our voting shares or their affiliated entities and certain of our executive officers and directors. Each preferred share identified in the following table converted into one common share upon the closing of the initial public offering of our Class A common shares.

Preferred share participant	Series A	Series B	Series C
5% or Greater Shareholders(1)			
Entities Managed by Baker Bros. Advisors LLP	16,057,618	3,598,392	4,155,000
HH RSV-XVII Holdings Limited	—	1,151,485	3,196,154
ArrowMark Funds	—	575,691	447,458
Cormorant Funds	—	—	830,999
Deerfield Special Situations Fund, L.P	—	—	383,538
Entities Affiliated with Dr. Robert Desnick	585,001	393,282	873,301
Sofinnova Venture Partners X L.P.	—	—	894,943
Officers and Directors(2)			
Sanj K. Patel	428,203	71,967	63,922
Stephen Mahoney	107,050	—	6,392
Thomas Beetham	48,172	—	6,392
Chris Heberlig	74,935	—	6,392
Carsten Boess	107,050	—	6,392
Rasmus Holm-Jorgensen	48,172	—	6,392
Felix J. Baker(3)	16,057,618	3,598,392	4,155,000
Thomas R. Malley(4)	—	71,967	—

(1) Additional details regarding these shareholders and their equity holdings are provided in the section titled "Principal shareholders."

(2) Additional details regarding these officers' and directors' equity holdings are provided in the section titled "Principal shareholders."

(3) Dr. Baker is the beneficial owner of the preferred shares acquired by entities managed by Baker Bros. See "Principal shareholders" for additional details.

(4) Mr. Malley is the beneficial owner of the preferred shares acquired by Mossrock Capital, LLC. See "Principal shareholders" for additional details.

Our directors, Felix J. Baker, Ph.D. and Stephen R. Biggar, M.D., Ph.D., are associated with 667, L.P. and Baker Brothers Life Sciences, L.P., which beneficially own more than 5% of our Class A common shares.

Investors' rights agreement

In connection with our Series C preferred share financing, we entered into an amended and restated investors' rights agreement, or the investors' rights agreement, with holders of our preferred shares, including certain executive officers, holders of 5% of any class of our voting shares and entities affiliated with certain of our directors. The investors' rights agreement, among other things, grants these shareholders specified registration rights with respect to Class A common shares held by them, including common shares issued or issuable upon conversion of any other class of our common shares convertible into, or options, warrants or other securities exercisable for, our Class A common shares held by them. For more information regarding the registration rights provided in these agreements, please refer to the section entitled "Description of share capital—Registration rights."

Indemnification agreements

We have entered into indemnification agreements with all of our directors and executive officers. These agreements, among other things, require us or will require us to indemnify each director (and in certain cases their related investment funds) and executive officer to the fullest extent permitted by Bermuda law, including indemnification of expenses such as attorneys' fees, judgments, fines and settlement amounts incurred by the director or executive officer in any action or proceeding, including any action or proceeding by or in right of our company, arising out of such person's services as a director or executive officer.

Employment agreements

We have entered into employment agreements with our named executive officers. The material terms of those arrangements are described in "Executive and director compensation—Executive compensation arrangements." In addition, we have entered into employment agreements with certain of our officers as described below.

Chris Heberlig

Chris Heberlig serves as our Executive Vice President and Chief Financial Officer. Mr. Heberlig's employment agreement provides for an annual base salary of $350,000, which may be changed from time to time in the discretion of our board of directors. Mr. Heberlig is eligible to earn an annual discretionary cash bonus with a target of 35% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Thomas Beetham

Thomas Beetham serves as our Executive Vice President and Chief Legal Officer. Mr. Beetham's employment agreement provides for an annual base salary of $375,000, which may be changed from time to time in the discretion of our board of directors. Mr. Beetham is eligible to earn an annual discretionary cash bonus with a target of 35% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Carsten Boess

Carsten Boess serves as our Executive Vice President, Corporate Affairs. Mr. Boess' employment agreement provides for an annual base salary of $400,000, which may be changed from time to time in the discretion of our board of directors. Mr. Boess is eligible to earn an annual discretionary cash bonus with a target of 35% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Rasmus Holm-Jorgensen

Rasmus Holm-Jorgensen serves as our Senior Vice President, Chief Strategy and Portfolio Officer. Mr. Holm-Jorgensen's employment agreement provides for an annual base salary of $337,740, which may be changed from time to time in the discretion of our board of directors. Mr. Holm-Jorgensen is eligible to earn an annual discretionary cash bonus with a target of 30% of base salary based on our board of directors' assessment of his individual performance as well as overall company performance.

Share option grants to officers and directors

We have granted share options for our Class A common shares to our named executive officers and certain of our directors as more fully described in the section entitled "Executive and director compensation." In December 2015, we granted 125,941 share options each to Messrs. Heberlig, Beetham and Boess and 75,565 share options to Mr. Holm-Jorgensen, each at an exercise price of $1.59 per share. In June 2017, we granted 62,475 share options to Mr. Heberlig, 66,135 share options to Mr. Beetham, 36,598 share options to Mr. Boess and 40,689 share options to Mr. Holm-Jorgensen, each at an exercise price of $3.80. In March 2018, we granted 109,795 share options each to Messrs. Heberlig and Beetham, 9,149 share options to Mr. Boess, and 36,598 share options to Mr. Holm-Jorgensen, each at an exercise price of $10.36. In September 2018, we granted 45,000 share options each to Messrs. Heberlig and Beetham, 20,000 share options to Mr. Boess, and 15,000 share options to Mr. Holm-Jorgensen, each at an exercise price of $30.93.

Policies and procedures for related person transactions

Our board of directors adopted a written related person transaction policy setting forth the policies and procedures for the review and approval or ratification of related person transactions. This policy covers, with certain exceptions set forth in Item 404 of Regulation S-K under the Securities Act, any transaction, arrangement or relationship, or any series of similar transactions, arrangements or relationships, in which we were or are to be a participant, where the amount involved exceeds $120,000 in any fiscal year and a related person had, has or will have a direct or indirect material interest, including without limitation, purchases of goods or services by or from the related person or entities in which the related person has a material interest, indebtedness, guarantees of indebtedness and employment by us of a related person. In reviewing and approving any such transactions, our audit committee considers all relevant facts and circumstances, including, but not limited to, whether the transaction is on terms comparable to those that could be obtained in an arm's length transaction and the extent of the related person's interest in the transaction.

Principal shareholders

The following table sets forth information with respect to the beneficial ownership of our Class A common shares, Class A1 common shares, Class B common shares and Class B1 common shares, as of December 31, 2018, and as adjusted to reflect the sale of Class A common shares in this offering and Class A1 common shares in the concurrent private placement, by:

- each person or group of affiliated persons known by us to beneficially own more than 5% of our Class A common shares or Class B common shares;
- each of our named executive officers and directors; and
- all of our executive officers and directors as a group.

The number of shares beneficially owned by each shareholder is determined under rules issued by the SEC. Under these rules, beneficial ownership includes any shares over which the individual or entity has sole or shared voting power or investment power. Applicable percentage ownership is based on 15,797,220 Class A common shares outstanding, 4,638,855 Class B common shares outstanding, 12,995,954 Class A1 common shares outstanding, and 16,057,618 Class B1 common shares outstanding, each as of December 31, 2018. In computing the number of shares beneficially owned by an individual or entity and the percentage ownership of that person, common shares subject to options or other rights held by such person that are currently exercisable or will become exercisable within 60 days of December 31, 2018 are considered outstanding, although these shares are not considered outstanding for purposes of computing the percentage ownership of any other person.

Unless noted otherwise, the address of all listed shareholders is c/o Kiniksa Pharmaceuticals Corp., 100 Hayden Avenue, Lexington, Massachusetts 02421. Each of the shareholders listed has sole voting and investment power with respect to the shares beneficially owned by the shareholder unless noted otherwise, subject to community property laws where applicable.

Our Class B common shares are convertible into Class A common shares or Class B1 common shares at any time at the option of the holder, with prior notice to us, on a one-for-one basis. Accordingly, each holder of Class B common shares is deemed to be the beneficial owner of, in each case, an equal number of Class A common shares and Class B1 common shares, in addition to any other Class A common shares or Class B1 common shares beneficially owned by such holder.

Our Class A1 common shares are convertible into Class A common shares at any time at the option of the holder, with prior notice to us, on a one-for-one basis, unless, 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. Accordingly, each holder of Class A1 common shares is deemed to be the beneficial owner of our number of Class A common shares that would result in such holder owning up to 4.99% of the issued and outstanding Class A common shares, in addition to any other Class A common shares beneficially owned by such holder.

Our Class B1 common shares are convertible into Class A common shares or Class B common shares at any time at the option of the holder, with prior notice to us, on a one-for-one basis, unless, 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. Accordingly, each holder of Class B1 common shares is deemed to be the beneficial owner of the number of Class A common shares and Class B common shares, in each case, that would result in such holder owning up to 4.99% of our issued and outstanding Class A common shares, in addition to any other Class A common shares or Class B common shares beneficially owned by such holder.

	Beneficial ownership before the offering and concurrent private placement								% of total voting power before the offering and concurrent private placement	Beneficial ownership after the offering and concurrent private placement								% to vot pow af offer a concurr priv placem
	Class A common shares		Class A 1 common shares		Class B common shares		Class B1 common shares			Class A common shares		Class A 1 common shares		Class B common shares		Class B1 common shares		
	Shares	%	Shares	%	Shares	%	Shares	%		Shares	%	Shares	%	Shares	%	Shares	%	
5% Shareholders																		
Arrowmark Funds(1)	1,020,408	6.46%	—	—	—	—	—	—	1.64%	1,023,149	5.54%	—	—	—	—	—	—	1.5
Sofinnova Venture Partners X, L.P.(2)	894,923	5.67%	—	—	—	—	—	—	1.44%	894,923	4.85%	—	—	—	—	—	—	1.3
Cormorant Funds(3)	830,999	5.26%	—	—	—	—	—	—	1.34%	830,999	4.50%	—	—	—	—	—	—	1.2
Entities Affiliated with Dr. Robert Desnick(4)	932,851	5.83%	—	—	214,101	4.62%	214,101	1.32%	4.94%	715,091	3.83%	—	—	214,101	4.62%	214,101	1.32%	4.4
Deerfield Special Situations Fund, L.P.(5)	1,226,890	7.78%	—	—	—	—	—	—	1.97%	1,226,890	6.65%	—	—	—	—	—	—	1.8
HH RSV-XVII Holdings Limited(6)	1,388,888	8.79%	4,347,639	33.45%	—	—	—	—	2.23%	1,388,888	7.53%	4,347,639	29.0%	—	—	—	—	2.1
Entities managed by Baker Bros. Advisors LP(7)	3,028,140	19.14%	7,753,392	59.66%	—	—	16,057,618	100%	4.87%	3,028,140	16.39%	9,753,392	65.0%	—	—	16,057,618	100%	4.6
Officers and Directors																		
Sanj K. Patel(8)	2,508,498	13.97%	—	—	1,526,160	32.90%	1,526,160	8.68%	25.12%	2,508,498	12.17%	—	—	1,526,160	32.90%	1,526,160	8.68%	24.0
Thomas Beetham(9)	651,202	3.98%	—	—	414,157	8.93%	414,157	2.51%	6.79%	651,202	3.42%	—	—	414,157	8.93%	414,157	2.51%	6.5
Carsten Boess(10)	682,133	4.16%	—	—	473,035	10.20%	473,035	2.86%	7.74%	682,133	3.58%	—	—	473,035	10.20%	473,035	2.86%	7.4
Chris Heberlig(11)	676,500	4.13%	—	—	440,920	9.50%	440,920	2.67%	7.22%	676,500	3.55%	—	—	440,920	9.50%	440,920	2.67%	6.9
Rasmus Holm-Jorgensen(12)	496,411	3.06%	—	—	340,960	7.35%	340,960	2.08%	5.59%	496,411	2.63%	—	—	340,960	7.35%	340,960	2.08%	5.3
Stephen Mahoney(13)	942,131	5.67%	—	—	656,027	14.14%	656,027	3.93%	10.74%	942,131	4.89%	—	—	656,027	14.14%	656,027	3.93%	10.3
John F. Paolini, M.D., Ph.D. (14)	207,723	1.30%	—	—	—	—	—	—	—	207,723	1.11%	—	—	—	—	—	—	
Felix J. Baker, Ph.D.(7)	3,028,140	19.14%	7,753,392	59.66%	—	—	16,057,618	100%	4.87%	3,028,140	16.39%	9,753,392	65.0%	—	—	16,057,618	100%	4.6
Stephen R. Biggar, M.D., Ph.D.(15)	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	
Thomas R. Malley(16)	114,167	*	—	—	—	—	—	—	*	114,167	*	—	—	—	—	—	—	
Tracey L. McCain(17)	15,096	*	—	—	—	—	—	—	—	15,096	*	—	—	—	—	—	—	
Kimberly J. Popovits(18)	15,096	*	—	—	—	—	—	—	—	15,096	*	—	—	—	—	—	—	
Barry D. Quart, Pham. D.(19)	48,605	*	—	—	—	—	—	—	—	48,605	*	—	—	—	—	—	—	
All executive officers and directors as a group (11 persons)(20)	8,176,966	48.19%	7,753,392	59.66%	3,037,264	65.47%	19,094,882	100%	54.74%	8,176,966	41.54%	9,735,392	65.0%	3,037,264	65.47%	19,094,882	100%	52.4

* Represents beneficial ownership less than 1%

(1) Consists of (a) 173,010 Class A common shares held directly by ArrowMark Fundamental Opportunity Fund, L.P. ("Opportunity Fund"), (b) 289,127 Class A common shares held directly by Meridian Small Cap Growth Fund ("Meridian"), (c) 224,726 Class A common shares held directly by Meridian Growth Fund ("Meridian Growth"), (d) 52,034 Class A common shares held directly by Lookfar Investments LLC ("Lookfar"), (e) 93,490 Class A common shares held directly by Iron Horse Investments LLC ("Iron Horse"), (f) 185,279 Class A common shares held directly by THB Iron Rose LLC ("Rose"), and (g) 2,741 Class A common shares held directly by THB Iron Rose LLC, Life Science Portfolio ("Life Science") (collectively, the "ArrowMark Funds"). ArrowMark Partners GP, LLC ("ArrowMark GP") is the general partner of Opportunity Fund and David Corkins is the managing member of ArrowMark GP. ArrowMark Colorado Holdings LLC ("ArrowMark Colorado") is investment advisor to Meridian, Meridian Growth, Lookfar, Iron Horse, Rose and Life Science (collectively, the "ArrowMark Colorado Funds"). Mr. Corkins is a managing member of ArrowMark Colorado and Mr. Yao is a portfolio manager of ArrowMark Colorado. Mr. Corkins may be considered the beneficial owner of the shares held by the ArrowMark Funds and Mr. Yao may be considered the beneficial owner of the shares held by ArrowMark Colorado. The principal business address of the ArrowMark Funds is 100 Fillmore Street, Suite 325, Denver, Colorado 80206. The foregoing is based on information known to us.

(2) Sofinnova Management IX, L.L.C. ("Sofinnova Management") is the general partner of Sofinnova Venture Partners X, L.P. ("Sofinnova X") and Anand Mehra, Michael Powell and James Healy are the managing members of Sofinnova Management. Sofinnova Management, Anand Mehra, Michael Powell and James Healy may be deemed to beneficially own the shares owned by Sofinnova X. Such entities and individuals disclaim beneficial ownership over all shares except to the extent of any pecuniary interest therein. The address for Sofinnova X and Sofinnova Management is 3000 Sand Hill Road, Building 4, Suite 250, Menlo Park, California 94025. The foregoing is based on information known to us.

(3) Consists of (a) 640,617 Class A common shares held directly by Cormorant Private Healthcare Fund I, L.P. ("Cormorant I"), (b) 161,962 Class A common shares held directly by Cormorant Global Healthcare Master Fund, LP ("Cormorant Master Fund"); and (c) 28,420 Class A common shares held directly by CRMA SPV, L.P. ("CRMA" and with Cormorant I and Cormorant Master Fund, the "Cormorant Funds"). Cormorant Global Healthcare GP, LLC ("Global GP") is the general partner of Cormorant Master Fund and Cormorant Private Healthcare GP, LLC ("Private GP") is the general partner of Cormorant I. Bihua Chen serves as the managing member of both Global GP and Private GP. Cormorant Asset Management LP ("Cormorant Management") serves as the investment manager to Cormorant I, Cormorant Master Fund and CRMA, and Ms. Chen serves as the managing member of Cormorant Asset Management. Ms. Chen may be deemed to beneficially own the shares held by the Cormorant Funds. The address of the Cormorant Funds, Global GP, Private GP, Cormorant Management, and Ms. Chen is 200 Clarendon Street, 52nd Floor, Boston, MA 02116. The foregoing information is based on a Schedule 13G filed on June 4, 2018 and information known to us.

(4) Consists of (a) 477,439 Class A common shares held by Edward Schuchman, Ph.D., as the trustee for the Desnick / Herzig 2012 GST Trust UAD 10/23/12 ("Trust") and New Direction IRA, Inc. FBO Robert Desnick Roth IRA ("New Direction"), (b) 214,101 Class B common shares held directly by New Direction, (c) 3,659 Class A common shares that Dr. Desnick has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options and (d) 237,652 Class A common shares directly held by Dr. Desnick. Dr. Desnick directs the voting and investment of the shares held by Trust and New Direction and may be deemed to beneficially own the shares owned by Trust and New Direction. The foregoing information is based on a Schedule 13G filed on July 19, 2018 and information known to us.

(5) Deerfield Mgmt, L.P. is the general partner of Deerfield Special Situations Fund, L.P. (the "Fund"). Deerfield Management Company, L.P. is the investment manager of the Fund. James E. Flynn is the sole member of the general partner of each of Deerfield Mgmt, L.P. and Deerfield Management Company, L.P. Deerfield Mgmt, L.P., Deerfield Management and Mr. Flynn may be deemed to be the beneficial owners of the shares owned by Fund. Deerfield Mgmt, L.P., Deerfield Management Company, L.P. and Mr. Flynn disclaim beneficial ownership of such shares except to the extent of its or his pecuniary interest therein. The address for the Fund is 780 Third Avenue, 37th Floor, New York, NY 10017. The foregoing information is based on a Schedule 13G filed on May 31, 2018 and information known to us.

(6) Consists of (a) 1,388,888 Class A common shares and (b) 4,347,639 Class A1 common shares held directly by HH RSV-XVII Holdings Limited ("HH RSV-XVII"). Hillhouse Fund III, L.P. ("Fund III") is the sole owner of HH RSV-XVII and Hillhouse Capital Management, Ltd. ("Hillhouse Capital Management") acts as the sole management company of Fund III. Wei Cao is the managing director of Hillhouse Capital Management. As a result, Fund III, Hillhouse Capital Management and Wei Cao may be considered beneficial owners of the shares held by HH RSV-XVII. The address of HH RSV-XVII is Walkers Corporate Limited, Cayman Corporate Centre, 27 Hospital Road, Grand Cayman, KYI-9008, Cayman Islands. The foregoing is based on information known to us.

(7) Beneficial ownership before the offering and the concurrent private placement consists of (a) 2,700,597 Class A common shares held directly by Baker Brothers Life Sciences, L.P. ("BBLS"), (b) 299,403 Class A common shares held by 667, L.P. ("667" and with BBLS, the "Baker Funds") (c) 7,018,874 Class A1 common shares held directly by BBLS, (d) 734,581 Class A1 common shares held by 667, (e) 14,658,102 Class B1 common shares held directly by BBLS, (f) 1,399,516 Class B1 common shares held directly by 667, (g) 14,070 Class A common shares that Felix J. Baker (a member of our board) has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options, and (h) 14,070 Class A common shares that Stephen R. Biggar (a member of our board) has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options. Beneficial ownership after the offering and the concurrent private placement gives effect to the sale and issuance of 2,000,000 Class A1 common shares in the concurrent private placement. Baker Bros. Advisors LP ("Advisors") is the management company and investment advisor to BBLS and 667. Dr. Baker is a managing member of Advisors and may be deemed to beneficially own the shares owned by the Baker Funds. Dr. Baker disclaims beneficial ownership of such shares except to the extent of any pecuniary interest therein. The address for the Baker Funds is 860 Washington Street, 3rd Floor, New York, NY 10014. The foregoing information is based on a Schedule 13D filed on May 31, 2018 and information known to us.

(8) Consists of (a) 245,685 Class A common shares held directly by Mr. Patel and 109,795 Class A common shares held by Mr. Patel as trustee for the Manisha S. Patel 2016 Irrevocable Trust, (b) 1,526,160 Class B common shares, and (c) 626,858 Class A common shares that Mr. Patel has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(9) Consists of (a) 79,589 Class A common shares, (b) 414,157 Class B common shares and (c) 157,456 Class A common shares that Mr. Beetham has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(10) Consists of (a) 79,589 Class A common shares, (b) 473,035 Class B common shares, and (c) 129,509 Class A common shares that Mr. Boess has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(11) Consists of (a) 42,991 Class A common shares held directly by Mr. Heberlig and 36,598 Class A common shares held by Sandra C. Heberlig, Mr. Heberlig's spouse, as trustee for the Christopher J. Heberlig 2017 Irrevocable Trust, (b) 440,920 Class B common shares, and (c) 155,991 Class A common shares that Mr. Heberlig has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(12) Consists of (a) 64,949 Class A common shares, (b) 340,960 Class B common shares, and (c) 90,502 Class A common shares that Mr. Holm-Jorgensen has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(13) Consists of (a) 79,589 Class A common shares held directly by Mr. Mahoney and 36,598 Class A common shares held by Krisha S. Mahoney, Mr. Mahoney's spouse, as trustee for the Stephen F. Mahoney 2016 Irrevocable Trust, (b) 656,027 Class B common shares, and (c) 169,917 Class A common shares that Mr. Mahoney has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(14) Consists of 207,723 Class A common shares that Dr. Paolini has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(15) Dr. Biggar has the right to acquire 14,070 Class A common shares within 60 days following December 31, 2018 pursuant to the exercise of share options. However, the policy of the Baker Funds and the Advisors does not permit Dr. Biggar to receive compensation for serving on our board of directors, and the Baker Funds are instead entitled to the pecuniary interest in any compensation received for his service. Therefore, Dr. Biggar has no voting or dispositive power and no pecuniary interest in these share options.

(16) Consists of (a) 71,967 Class A common shares held by Mossrock Capital, LLC ("Mossrock") and (b) 42,200 Class A common shares that Mr. Malley has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options. Mr. Malley is the president of Mossrock and may be deemed to beneficially own the shares owned by Mossrock. The address of Mossrock is 19 Martin Lane, Englewood, CO 80113.

(17) Includes 15,096 Class A common shares that Ms. McCain has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(18) Includes 15,096 Class A common shares that Ms. Popovits has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(19) Includes 48,605 Class A common shares that Dr. Quart has the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

(20) Consists of (a) 702,812 Class A common shares, (b) 7,753,392 Class A1 common shares, (c) 3,037,264 Class B common shares, (d) 16,057,618 Class B1 common shares, and (e) 1,469,598 Class A common shares that all executive officers and directors as a group have the right to acquire within 60 days following December 31, 2018 pursuant to the exercise of share options.

Description of share capital

The following description of our share capital and provisions of our memorandum of association and amended and restated bye-laws are summaries. You should also refer to our memorandum of association and amended and restated bye-laws, which are filed as exhibits to the registration statement of which this prospectus is part.

General

We are an exempted company incorporated under the laws of Bermuda. We are registered with the Registrar of Companies in Bermuda under registration number 50484. We were incorporated on July 21, 2015. Our registered office is located in Bermuda at Clarendon House, 2 Church Street, Hamilton HM11, Bermuda.

The objects of our business are unrestricted, and we have the capacity of a natural person. We can therefore undertake activities without restriction on our capacity.

Since our incorporation, other than a subdivision of our designated and issued share capital, there have been no material changes to our share capital, mergers, amalgamations or consolidations of us or any of our subsidiaries, no material changes in the mode of conducting our business, no material changes in the types of products produced or services rendered. There have been no bankruptcy, receivership or similar proceedings with respect to us or our subsidiaries.

There have been no public takeover offers by third parties for our shares nor any public takeover offers by us for the shares of another company that have occurred during the last or current financial years.

Share capital

Following this offering and the concurrent private placement, our authorized share capital will consist of 200,000,000 shares, par value $0.000273235 per share, and there will be 18,452,204 Class A common shares, 4,638,855 Class B common shares, 14,995,954 Class A1 common shares and 16,057,618 Class B1 common shares issued and outstanding. As of December 31, 2018, there were 41 holders of record of our Class A common shares, 12 holders of record of our Class B common shares, four holders of record of our Class A1 common shares and two holders of record of our Class B1 common shares. The actual number of shareholders of our Class A common shares is greater than this number of record holders, and includes shareholders who are beneficial owners, but whose Class A common shares are held in street name by brokers and other nominees. This number of holders of record also does not include shareholders whose Class A common shares may be held in trust by other entities.

Pursuant to our amended and restated bye-laws, subject to the requirements of Nasdaq and subject to any resolution of the shareholders to the contrary, our board of directors is authorized to issue any of our designated but unissued shares. There are no limitations on the right of non-Bermudians or non-residents of Bermuda to hold or vote our shares.

Common shares

We have four classes of shares: Class A, Class B, Class A1 and Class B1. Class A and Class B common shares are voting common shares, or together the voting common shares, and Class A1 and Class B1 are non-voting common shares. Except as described in this prospectus with respect to voting rights conversion and transferability each common share has the same rights and powers of, rank equally to, share ratably

with and are identical in all respects and as to all matters with each other common share. In the event of our liquidation, dissolution or winding up, the holders of our common shares are entitled to share equally and ratably in our assets, if any, remaining after the payment of all of our debts and liabilities, subject to any liquidation preference on any issued and outstanding preferred shares. None of our common shares have pre-emptive, redemption or sinking fund rights.

Class A common shares

The shares being offered in this offering are our Class A common shares. As of December 31, 2018, there were 15,797,220 Class A common shares issued and outstanding. All Class A common shares are fully paid and non-assessable.

Class B common shares

As of December 31, 2018, there were 4,638,855 Class B common shares outstanding. Each holder of Class B common shares may convert any portion of its Class B common shares into Class A common shares or Class B1 common shares at any time. In addition, each Class B common share automatically converts into one Class A common share upon transfer, except for transfers to or between affiliated holders. Our Class B common shares also have greater voting power than our Class A common shares, as described in "—Voting rights."

Class A1 common shares

As of December 31, 2018, there were 12,995,954 Class A1 common shares issued and outstanding. Each holder of Class A1 common shares may elect to convert any portion of its Class A1 common shares into voting Class A common shares at any time, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own or would beneficially own more than 4.99% of the issued and outstanding Class A common shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act. A holder of Class A1 common shares may increase, decrease or waive this limitation on ownership by providing us with 61-days' notice.

Class B1 common shares

As of December 31, 2018, there were 16,057,618 Class B1 common shares issued and outstanding. Each holder of Class B1 common shares may elect to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time, unless, immediately prior to or following such conversion, the holder and its affiliates beneficially own or would beneficially own more than 4.99% of the issued and outstanding Class A common shares or any other class of equity security (other than an exempted security) that is registered pursuant to Section 12 of the Exchange Act. A holder of Class B1 common shares may increase, decrease or waive this limitation on ownership by providing us with 61-days' notice. In addition, each Class B1 common share automatically converts into one Class A common share upon transfer, except for transfers to or between affiliated holders.

Preferred shares

Under Bermuda law and our amended and restated bye-laws, our board of directors is authorized to issue preferred shares in one or more series without shareholder approval. Our board of directors has the discretion under the bye-laws to determine the rights, preferences, privileges and restrictions, including voting rights, dividend rights, conversion rights, redemption privileges and liquidation preferences, of each series of preferred shares, without any further shareholder approval. The rights with respect to a series of preferred shares may be greater than the rights attached to our Class A common shares. It is not possible

to state the actual effect of the issuance of any preferred shares on the rights of holders of our common shares until our board of directors determines the specific rights attached to those preferred shares. The effect of issuing preferred shares could include, among other things, one or more of the following:

- restricting dividends in respect of our Class A common shares;
- diluting the voting power of our Class A common shares or providing that holders of preferred shares have the right to vote on matters as a class;
- impairing the liquidation rights of our common shares; or
- delaying or preventing a change of control of us.

There are currently no preferred shares outstanding, and we have no present plans to issue any preferred shares following this offering and the concurrent private placement.

Voting rights

Unless a different majority is required by Bermuda law or by our amended and restated bye-laws, resolutions to be approved by holders of voting common shares require approval by a simple majority of votes cast at a meeting at which a quorum is present. Holders of our voting common shares vote together as a single class on all matters presented to the shareholders for their vote or approval, including the election of directors. Any individual who is a shareholder and who is present and entitled to vote at a meeting may vote in person, as may any corporate shareholder that is represented by a duly authorized representative at a meeting of shareholders. Our amended and restated bye-laws also permit attendance at general meetings by proxy, provided the instrument appointing the proxy is in the form specified in the bye-laws or such other form as the board of directors may determine.

Each Class A common share is entitled to one vote per share and each Class B common share is entitled to ten votes per share. Each Class A1 common share and Class B1 common share is non-voting. Immediately following this offering and the concurrent private placement, the holders of Class A common shares will account for 28.5% of our aggregate voting power and the holders of Class B common shares will account for the remaining 71.5% of our aggregate voting power. Our bye-laws generally provide that holders of our voting common shares are entitled to vote, on a non-cumulative basis, at all annual general and special general meetings of shareholders with respect to matters on which voting common shares are eligible to vote. However, these percentages may change depending on any conversion of Class A1 and Class B1 common shares into voting common shares, to the extent any are issued, and any conversion of Class B common shares into Class A common shares. See "—Common shares" for more information.

Dividend rights

Under Bermuda law and our amended and restated bye-laws, we may not declare or pay dividends if there are reasonable grounds for believing that: (i) we would after the payment be unable to pay our liabilities as they become due; or (ii) that the realizable value of our assets would thereby be less than our liabilities. Under our bye-laws, each common share is entitled to dividends if, as and when dividends are declared by our board of directors, subject to any preferred dividend right of the holders of any preferred shares. There are no restrictions on our ability to transfer funds (other than funds denominated in Bermuda dollars) in and out of Bermuda or to pay dividends to U.S. residents who are holders of our common shares.

We intend to retain future earnings, if any, to finance the operation and expansion of our business and do not anticipate paying any cash dividends on our Class A common shares in the foreseeable future. Any future determination related to our dividend policy will be made at the discretion of our board of directors after considering our financial condition, results of operations, capital requirements, business prospects and other factors the board of directors deems relevant.

We are a holding company and have no direct operations. As a result, we will depend upon distributions from our subsidiaries to pay any dividends.

Variation of rights

If at any time we have more than one class of shares, the rights attaching to any class, unless otherwise provided for by the terms of issue of the relevant class, may be varied either: (i) with the consent in writing of the holders of 75% of the issued shares of that class; or (ii) with the sanction of a resolution passed by a majority of the votes cast at a general meeting of the relevant class of shareholders at which a quorum consisting of at least two persons holding or representing one-third of the issued shares of the relevant class is present. Our amended and restated bye-laws specify that the creation or issue of shares ranking equally with existing shares or the purchase or redemption by us of our shares will not, unless expressly provided by the terms of issue of existing shares, vary the rights attached to existing shares. In addition, the creation or issue of preferred shares ranking prior to common shares will not be deemed to vary the rights attached to common shares or, subject to the terms of any other series of preferred shares, to vary the rights attached to any other series of preferred shares.

Transfer of shares

Our board of directors may in its absolute discretion and without assigning any reason refuse to register the transfer of a share that it is not fully paid. The board of directors may also refuse to recognize an instrument of transfer of a share unless it is accompanied by the relevant share certificate and such other evidence of the transferor's right to make the transfer as the board of directors shall reasonably require. Subject to these restrictions, a holder of common shares may transfer the title to all or any of such holder's common shares by completing a form of transfer in the form set out in the bye-laws (or as near thereto as circumstances admit) or in such other common form as the board of directors may accept. The instrument of transfer must be signed by the transferor and transferee, although in the case of a fully paid share the board of directors may accept the instrument signed only by the transferor.

Meetings of shareholders

Under the Companies Act, a company is required to convene at least one general meeting of shareholders each calendar year, which is referred to as the annual general meeting. However, the members may by resolution waive this requirement, either for a specific year or period of time, or indefinitely. When the requirement has been so waived, any member may, on notice to the company, terminate the waiver, in which case an annual general meeting must be called.

The Companies Act provides that a special general meeting of shareholders may be called by the board of directors of a company and must be called upon the request of shareholders holding not less than 10% of the paid-up capital of the company carrying the right to vote at general meetings. The Companies Act also requires that shareholders be given at least five days' advance notice of a general meeting, but the accidental omission to give notice to any person does not invalidate the proceedings at a meeting. Our bye-laws provide that our President or Chairman or any two directors or any director and secretary may

convene an annual general meeting or a special general meeting. Under our bye-laws, at least twenty days' notice of an annual general meeting or a special general meeting must be given to each shareholder entitled to vote at such meeting. This notice requirement is subject to the ability to hold such meetings on shorter notice if such notice is agreed: (i) in the case of an annual general meeting by all of the shareholders entitled to attend and vote at such meeting; or (ii) in the case of a special general meeting by a majority in number of the shareholders entitled to attend and vote at the meeting holding not less than 95% in nominal value of the shares entitled to vote at such meeting. The quorum required for a general meeting of shareholders is two or more persons present throughout the meeting and representing in person or by proxy a majority of the voting power of the issued and outstanding voting shares.

Access to books and records and dissemination of information

Members of the general public have a right to inspect the public documents of a company available at the office of the Registrar of Companies in Bermuda. These documents include a company's memorandum of association, including its objects and powers, and certain alterations to the memorandum of association. The shareholders have the additional right to inspect the bye-laws of a company, minutes of general meetings and a company's audited financial statements, which must be presented to the annual general meeting. The register of members of a company is also open to inspection by shareholders and by members of the general public without charge. The register of members is required to be open for inspection for not less than two hours in any business day (subject to the ability of a company to close the register of members for not more than thirty days in a year). A company is required to maintain its share register in Bermuda but may, subject to the provisions of the Companies Act, establish a branch register outside of Bermuda. A company is required to keep at its registered office a register of directors and officers that is open for inspection for not less than two hours in any business day by members of the public without charge. Bermuda law does not, however, provide a general right for shareholders to inspect or obtain copies of any other corporate records.

Election and removal of directors

Our amended and restated bye-laws provide that our board of directors shall consist of not less than five members and not more than such number of directors as the board of directors determine. Our board of directors currently consists of seven directors. Our board of directors is divided into three classes that are, as nearly as possible, of equal size. Each class of directors will be elected for a three-year term of office, but the terms will be staggered so that the term of only one class of directors expires at each annual general meeting. Following the election of all of the directors at the first general meeting of shareholders following the initial public offering of our Class A common shares, the initial terms of the Class I, Class II and Class III directors will expire in 2022, 2020 and 2021, respectively. At each succeeding annual general meeting, successors to the class of directors whose term expires at the annual general meeting will be elected for a three-year term.

A shareholder holding any percentage of the common shares in issue may propose for election as a director someone who is not an existing director or is not proposed by our board of directors. Where a director is to be elected at an annual general meeting, notice of any such proposal for election must be given not less than 90 days nor more than 120 days before the anniversary of the last annual general meeting prior to the giving of the notice or, in the event the annual general meeting is called for a date that is not less than 30 days before or after such anniversary the notice must be given not later than ten days following the earlier of the date on which notice of the annual general meeting was posted to shareholders or the date on which public disclosure of the date of the annual general meeting was made.

Where a director is to be elected at a special general meeting; provided, that our board of directors has determined that shareholders may nominate persons for election at such special general meeting, that notice must be given not later than seven days following the earlier of the date on which notice of the special general meeting was posted to shareholders or the date on which public disclosure of the date of the special general meeting was made.

A director may be removed, only with cause, by the shareholders, provided notice of the shareholders meeting convened to remove the director is given to the director. The notice must contain a statement of the intention to remove the director and a summary of the facts justifying the removal and must be served on the director not less than 14 days before the meeting. The director is entitled to attend the meeting and be heard on the motion for his removal.

Proceedings of board of directors

Our amended and restated bye-laws provide that our business is to be managed and conducted by our board of directors. Bermuda law permits individual and corporate directors, and there is no requirement in our amended and restated bye-laws or Bermuda law that directors hold any of our shares. There is also no requirement in our amended and restated bye-laws or under Bermuda law that our directors must retire at a certain age.

The remuneration of our directors is determined by the board of directors and each such director, other than directors who are employees of the Company, shall be paid a fee at a rate determined by the board of directors. The directors may also be paid all travel, hotel and other expenses properly incurred by them in connection with our business or their duties as directors.

A director who has a direct or indirect interest in any contract or arrangement with the Company must disclose such interest as required by the Companies Act. Such an interested director is not entitled to vote on or participate in any discussion in respect of any such contract or arrangement in which he or she is interested.

Indemnification of directors and officers

Section 98 of the Companies Act provides generally that a Bermuda company may indemnify its directors, officers and auditors against any liability which by virtue of any rule of law would otherwise be imposed on them in respect of any negligence, default, breach of duty or breach of trust, except in cases where such liability arises from fraud or dishonesty of which such director, officer or auditor may be guilty in relation to the Company. Section 98 further provides that a Bermuda company may indemnify its judgment is awarded in their favor or in which they are acquitted or granted relief by the Supreme Court of Bermuda pursuant to section 281 of the Companies Act.

We have adopted provisions in our amended and restated bye-laws that provide that we shall indemnify our officers and directors in respect of their actions and omissions, except in respect of their fraud or dishonesty. Our amended and restated bye-laws provide that the shareholders waive all claims or rights of action that they might have, individually or in right of the Company, against any of the Company's directors or officers for any act or failure to act in the performance of such director's or officer's duties, except in respect of any fraud or dishonesty of such director or officer. Section 98A of the Companies Act permits us to purchase and maintain insurance for the benefit of any officer or director in respect of any loss or liability attaching to him or her in respect of any negligence, default, breach of duty or breach of

trust, whether or not the Company may otherwise indemnify such officer or director. We will purchase and maintain a directors' and officers' liability policy for such a purpose.

Amendment of memorandum of association and bye-laws

Bermuda law provides that the memorandum of association of a company may be amended by a resolution passed at a general meeting of shareholders. Amendments to most provisions of our bye-laws require an affirmative vote of a majority of our board of directors and a majority of the issued and outstanding shares carrying the right to vote at general meetings at the relevant time. In addition, amendments to certain sections of our bye-laws containing anti-takeover provisions require an affirmative vote of at least 66% of the directors then in office and at least 66% of the voting power of the issued and outstanding shares carrying the right to vote at general meetings at the relevant time. These provisions make it more difficult for any person to remove or amend any provisions in our bye-laws that may have an anti-takeover effect.

Under Bermuda law, the holders of an aggregate of not less than 20% in par value of the Company's issued share capital or any class thereof have the right to apply to the Supreme Court of Bermuda for an annulment of any amendment of the memorandum of association adopted by shareholders at any general meeting, other than an amendment which alters or reduces a company's share capital as provided in the Companies Act. Where such an application is made, the amendment becomes effective only to the extent that it is confirmed by the Bermuda court. An application for an annulment of an amendment of the memorandum of association must be made within 21 days after the date on which the resolution altering the Company's memorandum of association is passed and may be made on behalf of persons entitled to make the application by one or more of their number as they may appoint in writing for the purpose. No application may be made by shareholders voting in favor of the amendment.

Amalgamations and mergers

The amalgamation or merger of a Bermuda company with another company or corporation (other than certain affiliated companies) requires the amalgamation or merger to be approved by the company's board of directors and by its shareholders. Unless the company's bye-laws provide otherwise, the approval of 75% of the shareholders voting at such meeting is required to approve the amalgamation or merger, and the quorum for such meeting must be two or more persons holding or representing more than one-third of the issued shares of the company. Our amended and restated bye-laws provide that the approval of the affirmative vote of a majority of votes cast at a meeting to approve the amalgamation or merger shall be sufficient, and the quorum for such meeting shall be two or more persons holding or representing a majority of the voting power of the issued and outstanding voting shares.

Under Bermuda law, in the event of an amalgamation or merger of a Bermuda company with another company or corporation, a shareholder of the Bermuda company who did not vote in favor of the amalgamation or merger and who is not satisfied that fair value has been offered for such shareholder's shares may, within one month of notice of the shareholders meeting, apply to the Supreme Court of Bermuda to appraise the fair value of those shares.

Business combinations

Although the Companies Act does not contain specific provisions regarding "business combinations" between companies organized under the laws of Bermuda and "interested shareholders," we have included these provisions in our bye-laws. Specifically, our bye-laws contain provisions which prohibit us from

engaging in a business combination with an interested shareholder for a period of three years after the date of the transaction in which the person became an interested shareholder, unless, in addition to any other approval that may be required by applicable law:

- prior to the date of the transaction that resulted in the shareholder becoming an interested shareholder, our board of directors approved either the business combination or the transaction that resulted in the shareholder becoming an interested shareholder;
- upon consummation of the transaction that resulted in the shareholder becoming an interested shareholder, the interested shareholder owned at least 85% of the voting power of our issued and outstanding voting shares at the time the transaction commenced; or
- after the date of the transaction that resulted in the shareholder becoming an interested shareholder, the business combination is approved by our board of directors and authorized at an annual or special general meeting of shareholders by the affirmative vote of at least $66^{2}/_{3}$% of the voting power of our issued and outstanding voting shares that are not owned by the interested shareholder.

For purposes of these provisions, a "business combination" includes recapitalizations, mergers, amalgamations, consolidations, exchanges, asset sales, leases, certain issues or transfers of shares or other securities and other transactions resulting in a financial benefit to the interested shareholder. An "interested shareholder" is any person or entity that beneficially owns 15% or more of our issued and outstanding voting shares and any person or entity affiliated with or controlling or controlled by that person or entity.

Shareholder suits

Class actions and derivative actions are generally not available to shareholders under Bermuda law. The Bermuda courts, however, would ordinarily be expected to permit a shareholder to commence an action in the name of a company to remedy a wrong to the company where the act complained of is alleged to be beyond the corporate power of the company or illegal, or would result in the violation of the company's memorandum of association or bye-laws. Furthermore, consideration would be given by a Bermuda court to acts that are alleged to constitute a fraud against the minority shareholders or, for instance, where an act requires the approval of a greater percentage of the company's shareholders than that which actually approved it.

When the affairs of a company are being conducted in a manner that is oppressive or prejudicial to the interests of some part of the shareholders, one or more shareholders may apply to the Supreme Court of Bermuda, which may make such order as it sees fit, including an order regulating the conduct of the company's affairs in the future or ordering the purchase of the shares of any shareholders by other shareholders or by the company.

Our amended and restated bye-laws contain a provision by virtue of which our shareholders waive any claim or right of action that they have, both individually and on our behalf, against any director or officer in relation to any action or failure to take action by such director or officer, except in respect of any fraud or dishonesty of such director or officer. We have been advised by the SEC that in the opinion of the SEC, the operation of this provision as a waiver of the right to sue for violations of federal securities laws would likely be unenforceable in U.S. courts.

Capitalization of profits and reserves

Pursuant to our amended and restated bye-laws, our board of directors may (1) capitalize any part of the amount of our share premium or other reserve accounts or any amount credited to our profit and loss account or otherwise available for distribution by applying such sum in paying up unissued shares to be allotted as fully paid bonus shares pro rata (except in connection with the conversion of shares) to the shareholders; or (2) capitalize any sum standing to the credit of a reserve account or sums otherwise available for dividend or distribution by paying up in full, partly paid or nil paid shares of those shareholders who would have been entitled to such sums if they were distributed by way of dividend or distribution.

Untraced shareholders

Our amended and restated bye-laws provide that our board of directors may forfeit any dividend or other monies payable in respect of any shares that remain unclaimed for six years from the date when such monies became due for payment. In addition, we are entitled to cease sending dividend warrants and checks by post or otherwise to a shareholder if such instruments have been returned undelivered to, or left uncashed by, such shareholder on at least two consecutive occasions or, following one such occasion, reasonable enquires have failed to establish the shareholder's new address. This entitlement ceases if the shareholder claims a dividend or cashes a dividend check or a warrant.

Certain provisions of Bermuda law

We have been designated by the Bermuda Monetary Authority as a non-resident for Bermuda exchange control purposes. This designation allows us to engage in transactions in currencies other than the Bermudan dollar, and there are no restrictions on our ability to transfer funds (other than funds denominated in Bermudan dollars) in and out of Bermuda or to pay dividends to U.S. residents who are holders of our common shares.

The Bermuda Monetary Authority has given its consent for the issue and free transferability of all of the common shares that are the subject of this offering to and between residents and non-residents of Bermuda for exchange control purposes, provided our shares remain listed on an appointed stock exchange, which includes the Nasdaq. Approvals or permissions given by the Bermuda Monetary Authority do not constitute a guarantee by the Bermuda Monetary Authority as to our performance or our creditworthiness. Accordingly, in giving such consent or permissions, neither the Bermuda Monetary Authority nor the Registrar of Companies in Bermuda shall be liable for the financial soundness, performance or default of our business or for the correctness of any opinions or statements expressed in this prospectus. Certain issues and transfers of common shares involving persons deemed resident in Bermuda for exchange control purposes require the specific consent of the Bermuda Monetary Authority. We have sought and have obtained a specific permission from the Bermuda Monetary Authority for the issue and transfer of our common shares up to the amount of our designated capital from time to time, and options, warrants, depository receipts, rights, loan notes, debt instruments and our other securities to persons resident and non-resident for exchange control purposes with the need for prior approval of such issue or transfer.

In accordance with Bermuda law, share certificates are only issued in the names of companies, partnerships or individuals. In the case of a shareholder acting in a special capacity (for example as a trustee), certificates may, at the request of the shareholder, record the capacity in which the shareholder

is acting. Notwithstanding such recording of any special capacity, we are not bound to investigate or see to the execution of any such trust.

Registration rights

Following the closing of the concurrent private placement, holders of 37,670,093 Class A common shares (including Class A common shares issuable upon the conversion of our Class A1 common shares, Class B common shares, and Class B1 common shares), which we refer to as registrable securities, or their transferees are entitled to the following rights with respect to the registration of such shares for public resale under the Securities Act pursuant to an amended and restated investors rights agreement by and among us and certain of our shareholders, until such shares can otherwise be sold without restriction under Rule 144, or until the rights otherwise terminate pursuant to the terms of the investors' rights agreement. The registration of our common shares as a result of the following rights being exercised would enable holders to trade these shares without restriction under the Securities Act when the applicable registration statement is declared effective.

If at any time the holders of a majority of the registrable securities request in writing that we effect a registration with respect to all or part of such registrable securities then outstanding, we may be required to register their shares. We are obligated to effect at most one registration in response to these demand registration rights. If the holders requesting registration intend to distribute their shares by means of an underwriting, the managing underwriter of such offering will have the right to limit the numbers of shares to be underwritten for reasons related to the marketing of the shares.

Piggyback registration rights

If at any time we propose to register any of our Class A common shares under the Securities Act, subject to certain exceptions, the holders of registrable securities are entitled to notice of the registration and to include their shares of registrable securities in the registration. If our proposed registration involves an underwriting, the managing underwriter of such offering will have the right to limit the number of shares to be underwritten for reasons related to the marketing of the shares.

Form S-3 registration rights

If, at any time after we become entitled under the Securities Act to register our shares on a registration statement on Form S-3, 25% of the holders of the registrable securities then outstanding on an as-converted into Class A common shares basis request in writing that we effect a registration with respect to registrable securities at an aggregate price to the public in the offering of at least $5.0 million, we will be required to effect such registration within 20 days after the date of such request; provided, however, that we will not be required to effect such a registration if, within any twelve-month period, we have already effected two registrations on Form S-3 for the holders of registrable securities. On the day we are eligible to use a Form S-3 registration statement, we are obligated to register any then outstanding registrable securities held by affiliates.

Expenses

Ordinarily, other than underwriting discounts and commissions, we will be required to pay all expenses incurred by us related to any registration effected pursuant to the exercise of these registration rights. These expenses may include all registration and filing fees, printing expenses, fees and disbursements of our counsel, reasonable fees and disbursements of a counsel for the selling securityholders and blue-sky fees and expenses.

Termination of registration rights

The registration rights terminate upon the earlier of the closing of a deemed liquidation event, as defined in our bye-laws, or, with respect to the registration rights of an individual holder, when the holder can sell all of such holder's registrable securities in a 90-day period without restriction under Rule 144 under the Securities Act.

Certain corporate anti-takeover provisions

Certain provisions in our amended and restated bye-laws may be deemed to have an anti-takeover effect and may delay, deter or prevent a tender offer or takeover attempt that a shareholder might consider to be in its best interests, including attempts that might result in a premium being paid over the market price for the Class A common shares. These provisions are also designed, in part, to encourage persons seeking to acquire control of us to first negotiate with our board of directors.

Preferred shares

Pursuant to our amended and restated bye-laws, preferred shares may be issued from time to time, and the board of directors is authorized to determine the rights, preferences, powers, qualifications, limitations and restrictions.

Classified board

In accordance with the terms of our bye-laws, our board of directors is divided into three classes, class I, class II and class III, with members of each class serving staggered three-year terms after their initial terms of office following their elections at the first general meeting of shareholders following our initial public offering of our Class A common shares. Our amended and restated bye-laws further provide that the authorized number of directors may be changed only by resolution of the board of directors. Any additional directorships resulting from an increase in the number of directors will be distributed among the three classes so that, as nearly as possible, each class will consist of one-third of the directors. Our classified board of directors could have the effect of delaying or discouraging an acquisition of us or a change in our management.

Removal of directors

In accordance with the terms of our amended and restated bye-laws, our directors may be removed only for cause by the affirmative vote of a majority of the votes entitled to be cast by our shareholders entitled to vote at an annual general election of directors. Any vacancy on our board, including a vacancy resulting from an enlargement of our board or from removal for cause not filled by the shareholders at the time, may be filled only by vote of a majority of our directors then in office.

Advance notice requirements for shareholder proposals and director nominations

Our amended and restated bye-laws provide that shareholders seeking to nominate candidates for election as directors or to bring business before an annual meeting of shareholders must provide timely notice of their proposal. Generally, to be timely, a shareholder's notice must be received at our principal executive offices not less than 90 days or more than 120 days prior to the first anniversary date of the last annual general meeting. Our amended and restated bye-laws also specify requirements as to the form and content of a shareholder's notice. These provisions may impede shareholders' ability to bring matters before an annual meeting of shareholders or make nominations for directors at an annual meeting of shareholders.

Choice of jurisdiction

Our amended and restated by-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 bye-laws, including any question regarding the existence and scope of any bye-law and/or whether there has been a breach of the Companies Act or the 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.

Amendment of certain bye-laws

Amendments to certain sections of our amended and restated bye-laws containing anti-takeover provisions will require an affirmative vote of at least 66% of the directors and at least 66% of the voting power of the issued and outstanding shares.

Registrar and transfer agent

A register of holders of the Class A common shares is maintained by Conyers Corporate Services (Bermuda) Limited in Bermuda, and a branch register is maintained in the United States by American Stock Transfer & Trust Company LLC, which also serves as transfer agent. The transfer agent's address is 6201 15th Avenue, Brooklyn, NY 11219.

Listing

Our Class A common shares are listed on The Nasdaq Global Select Market under the symbol "KNSA."

Material Bermuda and U.S. federal income tax considerations

The following is a discussion of the material Bermuda and U.S. federal income tax considerations that may be relevant to an investment decision by a potential investor with respect to our common shares.

Bermuda tax considerations

Taxation of the company

Under current Bermuda law, there is no income, corporate or profits tax or withholding tax, capital gains tax or capital transfer tax, estate or inheritance tax payable by us or our shareholders, other than shareholders ordinarily resident in Bermuda, if any. The Company has received from the Minister of Finance under The Exempted Undertaking Tax Protection Act 1966, as amended, an assurance that, in the event that Bermuda enacts legislation imposing tax computed on profits, income, any capital asset, gain or appreciation, or any tax in the nature of estate duty or inheritance, then the imposition of any such tax will not be applicable to the Company or to any of their operations or their shares, debentures or other obligations, until March 31, 2035. This assurance is subject to the proviso that it is not to be construed so as to prevent the application of any tax or duty to such persons as are ordinarily resident in Bermuda or to prevent the application of any tax payable in accordance with the provisions of the Land Tax Act 1967 or otherwise payable in relation to any property leased to the Company. The Company pays annual Bermuda government fees which fees are calculated on a sliding scale based on the assessable capital of the company. In addition, all entities employing individuals in Bermuda are required to pay a payroll tax and there are other sundry taxes payable, directly or indirectly, to the Bermuda government.

Taxation of shareholders

Currently, there is no Bermuda income, corporate or profits tax or withholding tax, capital gains tax or capital transfer tax, estate or inheritance tax payable by holders of our shares, other than shareholders ordinarily resident in Bermuda, if any.

Material U.S. federal income tax considerations to U.S. holders

The following discussion describes the material U.S. federal income tax consequences to U.S. Holders (as defined below) under present law of an investment in the Class A common shares. The effects of any applicable state or local laws, or other U.S. federal tax laws such as estate and gift tax laws, or the alternative minimum tax or the Medicare contribution tax on net investment income, are not discussed. This summary applies only to investors who acquire the Class A common shares in exchange for cash, hold the Class A common shares as capital assets within the meaning of Section 1221 of the Code (generally, property held for investment) and who have the U.S. dollar as their functional currency. This discussion is based on the Code, U.S. Treasury regulations promulgated thereunder, judicial decisions, published rulings and administrative pronouncements of the U.S. Internal Revenue Service, or the IRS, all as in effect as of the date of this prospectus. All of the foregoing authorities are subject to change, which change could apply retroactively and could affect the tax consequences described below.

The following discussion does not address all U.S. federal income tax consequences relevant to a holder's particular circumstances or to holders subject to particular rules, including, without limitation:

- U.S. expatriates and certain former citizens or long-term residents of the United States;
- persons whose functional currency is not the U.S. dollar;

- persons holding Class A common shares as part of a hedge, straddle or other risk reduction strategy or as part of a conversion transaction or other integrated investment;
- banks, insurance companies and other financial institutions;
- real estate investment trusts or regulated investment companies;
- brokers, dealers or traders in securities, commodities or currencies;
- S corporations or entities or arrangements treated as partnerships for U.S. federal income tax purposes;
- tax-exempt organizations or governmental organizations;
- individual retirement accounts or other tax deferred accounts;
- persons who acquired the Class A common shares pursuant to the exercise of any employee share option or otherwise as compensation;
- persons that own or are deemed to own 10% or more of our stock by vote or value directly, indirectly or constructively;
- persons subject to special tax accounting rules as a result of any item of gross income with respect to the Class A common shares being taken into account in an applicable financial statement;
- persons that hold Class A common shares through a permanent establishment or fixed base outside the United States; and
- persons deemed to sell Class A common shares under the constructive sale provisions of the Code.

We believe we are a "controlled foreign corporation" for U.S. federal income tax purposes, and therefore, if you are a U.S. shareholder owning 10% or more of our stock by vote or value directly, indirectly or constructively, the U.S. federal income tax consequences to you of owning our Class A common shares may be significantly different than those described below. If you own 10% or more of our stock by vote or value directly, indirectly or constructively, you should consult your tax advisors regarding the U.S. federal income tax consequence of your investment in our Class A common shares.

U.S. HOLDERS ARE URGED TO CONSULT THEIR TAX ADVISORS REGARDING THE APPLICATION OF THE U.S. FEDERAL TAX RULES TO THEIR PARTICULAR CIRCUMSTANCES AS WELL AS THE U.S. STATE AND LOCAL AND NON-U.S. TAX CONSEQUENCES TO THEM OF THE PURCHASE, OWNERSHIP AND DISPOSITION OF OUR CLASS A COMMON SHARES.

For purposes of this discussion, a "U.S. Holder" is a beneficial owner of Class A common shares that, for U.S. federal income tax purposes, is or is treated as any of the following:

- an individual who is a citizen or resident of the United States;
- a corporation (or another entity treated as a corporation for U.S. federal income tax purposes) created or organized under the laws of the United States, any state thereof, or the District of Columbia;
- an estate, the income of which is subject to U.S. federal income tax regardless of its source; or
- a trust that (1) is subject to the supervision of a U.S. court and the control of one or more "United States persons" (within the meaning of Section 7701(a)(30) of the Code), or (2) has a valid election in effect to be treated as a United States person for U.S. federal income tax purposes.

If you are an entity taxable as a partnership for U.S. federal income tax purposes that holds Class A common shares, your tax treatment generally will depend on your status and the activities of the partnership. Partnerships holding Class A common shares and the partners in such partnerships should consult their tax advisors regarding the U.S. federal income tax consequences applicable to them.

Taxation of dividends and other distributions on the Class A common shares

The discussion in this section "Taxation of dividends and other distributions on the Class A common shares" is subject to the discussion regarding passive foreign investment companies below. As discussed above under "Dividend policy," the Company does not currently intend to declare dividends on the Class A common shares in the foreseeable future. In the event the Company does pay dividends, the gross amount of any distribution to you with respect to the Class A common shares will be included in your gross income as dividend income when actually or constructively received to the extent that the distribution is paid out of our current or accumulated earnings and profits (as determined under U.S. federal income tax principles). To the extent the amount of the distribution exceeds our current and accumulated earnings and profits, it will be treated first as a return of your tax basis in the Class A common shares, and to the extent the amount of the distribution exceeds your tax basis, the excess will be taxed as capital gain. We do not intend to calculate our earnings and profits under U.S. federal income tax principles. Therefore, a U.S. Holder should expect that distributions will generally be reported as ordinary dividend income for such purposes. Dividends we pay will not be eligible for the dividends-received deduction available to corporations in respect of dividends received from U.S. corporations.

Subject to certain limitations, dividends paid by qualified foreign corporations to certain non-corporate U.S. Holders may be taxable at preferential tax rates. A non-U.S. corporation is generally treated as a qualified foreign corporation with respect to dividends paid on stock that is readily tradable on a securities market in the United States, such as Nasdaq, on which the Company has applied to list the Class A common shares. However, the preferential tax rates discussed above will not apply if we are treated as a passive foreign investment company with respect to the U.S. Holder for the taxable year in which a dividend is paid or the preceding year. Non-corporate U.S. Holders should consult their tax advisers regarding the availability of the reduced tax rate on dividends. Dividends will be included in a U.S. Holder's income on the date of the U.S. Holder's receipt of the dividend.

Dividends will generally constitute foreign source income for foreign tax credit limitation purposes. Any tax withheld with respect to distributions on the Class A common shares may, subject to a number of complex limitations, be claimed as a foreign tax credit against such U.S. Holder's U.S. federal income tax liability or may be claimed as a deduction for U.S. federal income tax purposes. The limitation on foreign taxes eligible for credit is calculated separately with respect to specific classes of income. For this purpose, dividends distributed by us with respect to the Class A common shares generally will constitute "passive category income." The rules with respect to the foreign tax credit are complex and may depend upon a U.S. Holder's particular circumstances. You should consult your tax advisor regarding the availability of the foreign tax credit under your particular circumstances.

Taxation of disposition of the Class A common shares

The discussion in this section "Taxation of disposition of Class A common shares" is subject to the discussion regarding passive foreign investment companies below. You will recognize gain or loss on any sale, exchange or other taxable disposition of Class A common shares equal to the difference between the amount realized (in U.S. dollars) on the disposition and your tax basis (in U.S. dollars) in the Class A common shares. Any such gain or loss will be capital gain or loss, and will be long-term capital gain or

loss if you have held the Class A common shares for more than one year at the time of the disposition. Otherwise, such gain or loss will be short-term capital gain or loss. Long-term capital gains recognized by certain non-corporate U.S. Holders, including individuals, generally will be taxable at reduced rates. The deductibility of capital losses is subject to limitations. Any such gain or loss you recognize generally will be treated as U.S. source income or loss for foreign tax credit limitation purposes. You should consult your tax advisor regarding the proper treatment of gain or loss in your particular circumstances.

Passive foreign investment company

Because we do not expect to earn revenue from our business operations during the current taxable year, and because our sole source of income currently is interest on bank accounts, we believe we will likely be a PFIC for our current taxable year. A non-U.S. corporation will be classified as a PFIC for any taxable year in which, after applying certain look-through rules, either:

- at least 75% of its gross income for such taxable year is passive income; or
- at least 50% of the value of its assets (based on an average of the quarterly values of the assets during a taxable year) is attributable to assets that produce or are held for the production of passive income.

For purposes of the above calculations, if a non-U.S. corporation owns, directly or indirectly, 25% or more of the total value of the outstanding shares of another corporation, it will be treated as if it (a) held a proportionate share of the assets of such other corporation and (b) received directly a proportionate share of the income of such other corporation. Passive income generally includes dividends, interest, rents, royalties and capital gains, but generally excludes rents and royalties which are derived in the active conduct of a trade or business and which are received from a person other than a related person.

A separate determination must be made each taxable year as to whether we are a PFIC (after the close of each such taxable year). Because the value of our assets for purposes of the asset test will generally be determined by reference to the market price of the Class A common shares, our PFIC status will depend in large part on the market price of the Class A common shares, which may fluctuate significantly. In addition, changes in the composition of our income or assets may cause us to become a PFIC.

If we are classified as a PFIC in any year with respect to which a U.S. Holder owns Class A common shares, we will continue to be treated as a PFIC with respect to such U.S. Holder in all succeeding years during which the U.S. Holder owns the Class A common shares, regardless of whether we continue to meet the tests described above unless we cease to be a PFIC and the U.S. Holder (1) has made a "deemed sale" election under the PFIC rules, (2) the U.S. Holder has a valid mark-to-market election in effect (as described below) or (3) the U.S. Holder makes a QEF Election (defined below) with respect to all taxable years in which we are a PFIC during such U.S. Holder's holding period in which we are a PFIC or makes a purging election to cause a deemed sale of the PFIC shares at their fair market value in conjunction with a QEF Election (see discussion below regarding such elections). If a U.S. Holder makes a deemed sale election, such U.S. Holder will be deemed to have sold the common shares held by such U.S. Holder at their fair market value, and any gain from such deemed sale would be subject to the rules described below. After the deemed sale election, so long as we do not become a PFIC in a subsequent taxable year, a U.S. Holder's Class A common shares subject to such election will not be treated as shares in a PFIC, and the rules described below with respect to any "excess distributions" or any gain from an actual sale or other disposition of the Class A common shares will not apply. U.S. Holders should consult their tax advisors as to the possibility and consequences of making a deemed sale election if we cease to be a PFIC.

For each taxable year we are treated as a PFIC with respect to you, you will be subject to special tax rules with respect to any "excess distribution" you receive and any gain you realize from a sale or other disposition (including a pledge) of Class A common shares, unless you (1) make a QEF Election (as defined below) with respect to all taxable years of your holding period during which we are a PFIC (as discussed below) or make a purging election to cause a deemed sale of the PFIC shares at their fair market value in conjunction with a QEF Election (see discussion below regarding such elections) or (2) make a mark-to-market election as discussed below. Distributions you receive in a taxable year that are greater than 125% of the average annual distributions you received during the shorter of the three preceding taxable years or your holding period for the Class A common shares will be treated as an excess distribution. Under these special tax rules, if you receive any excess distribution or realize any gain from a sale or other disposition of the Class A common shares:

- the excess distribution or gain will be allocated ratably over your holding period for the Class A common shares;
- the amount allocated to the current taxable year, and any taxable year before the first taxable year in which we were a PFIC, will be treated as ordinary income; and
- the amount allocated to each other year will be subject to the highest tax rate in effect for that year and an interest charge generally applicable to underpayments of tax will be imposed on the resulting tax attributable to each such year.

The tax liability for amounts allocated to years before the year of disposition or "excess distribution" cannot be offset by any net operating losses for such years, and gains (but not losses) realized on the sale of Class A common shares cannot be treated as capital, even if you hold the Class A common shares as capital assets.

If we are treated as a PFIC with respect to you for any taxable year, to the extent any of our subsidiaries are also PFICs, you will be deemed to own your proportionate share of any such lower-tier PFICs, and you may be subject to the rules described in the preceding two paragraphs with respect to the shares of such lower-tier PFICs you would be deemed to own. As a result, you may incur liability for any "excess distribution" described above if we receive a distribution from such lower-tier PFICs or if any shares in such lower-tier PFICs are disposed of (or deemed disposed of). You should consult your tax advisor regarding the application of the PFIC rules to any of our subsidiaries.

A U.S. Holder of "marketable stock" (as defined below) in a PFIC may make a mark-to-market election for such stock to elect out of the general tax treatment for PFICs discussed above. If you make a mark-to-market election for the Class A common shares, you will include in income for each year we are a PFIC an amount equal to the excess, if any, of the fair market value of the Class A common shares as of the close of your taxable year over your adjusted basis in such Class A common shares. Accordingly, a mark-to-market election may accelerate the recognition of income without a corresponding receipt of cash. You are allowed a deduction for the excess, if any, of the adjusted basis of the Class A common shares over their fair market value as of the close of the taxable year. However, deductions are allowable only to the extent of any net mark-to-market gains on the Class A common shares included in your income for prior taxable years. Amounts included in your income under a mark-to-market election, as well as gain on the actual sale or other disposition of Class A common shares, are treated as ordinary income. Ordinary loss treatment also applies to the deductible portion of any mark-to-market loss on Class A common shares, as well as to any loss realized on the actual sale or disposition of Class A common shares to the extent the amount of such loss does not exceed the net mark-to-market gains previously included for the

Class A common shares. Your basis in the Class A common shares will be adjusted to reflect any such income or loss amounts. If you make a valid mark-to-market election, the tax rules that apply to distributions by corporations which are not PFICs would apply to distributions by us, except the lower applicable tax rates for qualified dividend income would not apply. If we cease to be a PFIC when you have a mark-to-market election in effect, gain or loss realized by you on the sale of Class A common shares will be a capital gain or loss and taxed in the manner described above under "Taxation of disposition of the Class A common shares."

The mark-to-market election is available only for "marketable stock," which is stock that is traded in other than de minimis quantities on at least 15 days during each calendar quarter, or regularly traded, on a qualified exchange or other market, as defined in applicable U.S. Treasury regulations. Any trades that have as their principal purpose meeting this requirement will be disregarded. The Class A common shares are listed on The Nasdaq Global Select Market and, accordingly, provided the Class A common shares are regularly traded, if you are a holder of Class A common shares, the mark-to-market election would be available to you if we are a PFIC. Once made, the election cannot be revoked without the consent of the IRS unless the Class A common shares cease to be marketable stock. If we are a PFIC for any year in which a U.S. Holder owns Class A common shares but before a mark-to-market election is made, the interest charge rules described above will apply to any mark-to market gain recognized in the year the election is made. If any of our subsidiaries are or become PFICs, the mark-to-market election will not be available with respect to shares of such subsidiaries that are treated as owned by you. Consequently, you could be subject to the PFIC rules with respect to income of the lower-tier PFICs the value of which already had been taken into account indirectly via mark-to-market adjustments. You should consult your tax advisor as to the availability and desirability of a mark-to-market election, as well as the impact of such election on interests in any lower-tier PFICs.

Alternatively, a U.S. Holder can make an election, if we provide the necessary information, to treat us and each lower-tier PFIC (if any) as a qualified electing fund (a "QEF Election") in the first taxable year we (and any relevant subsidiaries) are treated as a PFIC with respect to the holder. If such election remains in place while we and any lower-tier PFIC subsidiaries are PFICs, we and our subsidiaries will not be treated as PFICs with respect to such U.S. Holder when we cease to be a PFIC. A U.S. Holder must make the QEF Election for each PFIC by attaching a separate properly completed IRS Form 8621 for each PFIC to the Holder's timely filed U.S. federal income tax return. We will provide the information necessary for a U.S. Holder to make a QEF Election with respect to us and will cause each lower-tier PFIC we control to provide such information.

If a U.S. Holder makes a QEF Election with respect to a PFIC, the Holder will be currently taxable on its pro rata share of the PFIC's ordinary earnings and net capital gain (at ordinary income and capital gain rates, respectively) for each taxable year that the entity is classified as a PFIC. Accordingly, a QEF election may accelerate the recognition of income without a corresponding receipt of cash. If a U.S. Holder makes a QEF Election with respect to us, any distributions paid by us out of our earnings and profits that were previously included in the Holder's income under the QEF Election would not be taxable to the holder. A U.S. Holder will increase its tax basis in its Class A common shares by an amount equal to any income included under the QEF Election and will decrease its tax basis by any amount distributed that is not included in the holder's income. In addition, a U.S. Holder will recognize capital gain or loss on the disposition of Class A common shares in an amount equal to the difference between the amount realized and the holder's adjusted tax basis in the Class A common shares. U.S. Holders should note that if they make QEF Elections with respect to us and any lower-tier PFICs, they may be required to pay U.S. federal income tax with respect to their Class A common shares for any taxable year significantly in excess of any

cash distributions received in such taxable year. U.S. Holders should consult their tax advisors regarding making QEF Elections in their particular circumstances.

If we are considered a PFIC, a U.S. Holder will also be subject to annual information reporting requirements. U.S. Holders should consult their tax advisors about the potential application of the PFIC rules to an investment in Class A common shares.

YOU ARE STRONGLY URGED TO CONSULT YOUR TAX ADVISOR REGARDING THE APPLICATION OF THE PFIC RULES TO YOUR INVESTMENT IN OUR CLASS A COMMON SHARES.

Information reporting and backup withholding

Dividend payments with respect to Class A common shares and proceeds from the sale, exchange or other disposition of Class A common shares may be subject to information reporting to the IRS and U.S. backup withholding. Certain U.S. Holders are exempt from backup withholding, including corporations and certain tax-exempt organizations. A U.S. Holder will be subject to backup withholding if such holder is not otherwise exempt and such holder:

- fails to furnish the holder's taxpayer identification number, which for an individual is ordinarily his or her social security number;
- furnishes an incorrect taxpayer identification number;
- is notified by the IRS that the holder previously failed to properly report payments of interest or dividends; or
- fails to certify under penalties of perjury that the holder has furnished a correct taxpayer identification number and that the IRS has not notified the holder that the holder is subject to backup withholding.

Backup withholding is not an additional tax. Any amounts withheld under the backup withholding rules may be allowed as a refund or a credit against the U.S. Holder's U.S. federal income tax liability, provided the required information is timely furnished to the IRS. U.S. Holders should consult their tax advisors regarding their qualification for an exemption from backup withholding and the procedures for obtaining such an exemption.

Additional reporting requirements

Certain U.S. Holders who are individuals (and certain entities) that hold an interest in "specified foreign financial assets" (which may include the Class A common shares) are required to report information relating to such assets, subject to certain exceptions (including an exception for Class A common shares held in accounts maintained by certain financial institutions). Penalties can apply if U.S. Holders fail to satisfy such reporting requirements. U.S. Holders should consult their tax advisors regarding the applicability of these requirements to their acquisition and ownership of Class A common shares.

Underwriting

The company and the underwriters named below have entered into an underwriting agreement with respect to the shares being offered in the offering. Subject to certain conditions, each underwriter has severally agreed to purchase the number of shares indicated in the following table. J.P. Morgan Securities LLC, Goldman Sachs & Co. LLC and Barclays Capital Inc. are the representatives of the underwriters.

Underwriters	Number of shares
J.P. Morgan Securities LLC	955,794
Goldman Sachs & Co. LLC	849,595
Barclays Capital Inc.	398,248
Wedbush Securities Inc.	238,949
JMP Securities LLC	212,398
Total	2,654,984

The underwriters will be committed to take and pay for all of the shares being offered in the offering, if any are taken, other than the shares covered by the option described below unless and until this option is exercised. The underwriting agreement also provides that if an underwriter defaults, the purchase commitments of non-defaulting underwriters may also be increased or the offering may be terminated.

The underwriters will have an option to buy up to an additional 398,247 Class A common shares from the company to cover sales by the underwriters of a greater number of shares than the total number set forth in the table above. They may exercise that option for 30 days from the date of this prospectus. If any shares are purchased pursuant to this option, the underwriters will severally purchase shares in approximately the same proportion as set forth in the table above.

The following table shows the per share and total underwriting discounts and commissions to be paid to the underwriters by the company. Such amounts are shown assuming both no exercise and full exercise of the underwriters' option to purchase 398,247 additional Class A common shares.

Paid by the company

	No exercise	Full exercise
Per Share	$ 1.00	$ 1.00
Total	$ 2,666,400	$ 3,066,360

Shares sold by the underwriters to the public will initially be offered at the public offering price set forth on the cover of this prospectus. Any shares sold by the underwriters to securities dealers may be sold at a discount of up to $0.60 per share from the public offering price. After the offering of the shares, the representatives may change the offering price and the other selling terms. The offering of the shares by the underwriters is subject to receipt and acceptance and subject to the underwriters' right to reject any order in whole or in part.

The company and its officers and directors have agreed or will agree with the underwriters, subject to certain exceptions, not to dispose of or hedge any of their common shares or securities convertible into or exchangeable for common shares during the period from the date of this prospectus continuing through

the date 90 days after the date of this prospectus, except with the prior written consent of the representatives. This agreement does not apply to any existing employee benefit plans.

Our Class A common shares are listed on The Nasdaq Global Select Market under the symbol "KNSA."

In connection with the offering, the underwriters may purchase and sell Class A common shares in the open market. These transactions may include short sales, stabilizing transactions and purchases to cover positions created by short sales. Short sales involve the sale by the underwriters of a greater number of shares than they are required to purchase in the offering, and a short position represents the amount of such sales that have not been covered by subsequent purchases. A "covered short position" is a short position that is not greater than the amount of additional Class A common shares for which the underwriters' option described above may be exercised. The underwriters may cover any covered short position by either exercising their option to purchase additional Class A common shares or purchasing shares in the open market. In determining the source of shares to cover the covered short position, the underwriters will consider, among other things, the price of shares available for purchase in the open market as compared to the price at which they may purchase additional Class A common shares pursuant to the option described above. "Naked" short sales are any short sales that create a short position greater than the amount of additional Class A common shares for which the option described above may be exercised. The underwriters must cover any such naked short position by purchasing shares in the open market. A naked short position is more likely to be created if the underwriters are concerned that there may be downward pressure on the price of the Class A common shares in the open market after pricing that could adversely affect investors who purchase in the offering. Stabilizing transactions consist of various bids for or purchases of Class A common shares made by the underwriters in the open market prior to the completion of the offering.

The underwriters may also impose a penalty bid. This occurs when a particular underwriter repays to the underwriters a portion of the underwriting discount received by it because the representatives have repurchased shares sold by or for the account of such underwriter in stabilizing or short covering transactions.

Purchases to cover a short position and stabilizing transactions, as well as other purchases by the underwriters for their own accounts, may have the effect of preventing or retarding a decline in the market price of the company's Class A common shares, and together with the imposition of the penalty bid, may stabilize, maintain or otherwise affect the market price of the common shares. As a result, the price of the Class A common shares may be higher than the price that otherwise might exist in the open market. The underwriters are not required to engage in these activities and may end any of these activities at any time. These transactions may be effected on Nasdaq, in the over-the-counter market or otherwise.

The company estimates that its share of the total expenses of the offering, excluding underwriting discounts and commissions, will be approximately $1.0 million. We will agree to reimburse the underwriters for expenses related to any applicable state securities filings and to the Financial Industry Regulatory Authority incurred by them in connection with this offering in an amount up to $20,000. In addition, the underwriters have agreed to reimburse us for certain expenses incurred by us in connection with this offering.

The company has agreed to indemnify the several underwriters against certain liabilities, including liabilities under the Securities Act of 1933.

The underwriters and their respective affiliates are full service financial institutions engaged in various activities, which may include sales and trading, commercial and investment banking, advisory, investment

management, investment research, principal investment, hedging, market making, brokerage and other financial and non-financial activities and services. Certain of the underwriters and their respective affiliates have provided, and may in the future provide, a variety of these services to the issuer and to persons and entities with relationships with the issuer, for which they received or will receive customary fees and expenses.

In the ordinary course of their various business activities, the underwriters and their respective affiliates, officers, directors and employees may purchase, sell or hold a broad array of investments and actively trade securities, derivatives, loans, commodities, currencies, credit default swaps and other financial instruments for their own account and for the accounts of their customers, and such investment and trading activities may involve or relate to assets, securities and/or instruments of the issuer (directly, as collateral securing other obligations or otherwise) and/or persons and entities with relationships with the issuer. The underwriters and their respective affiliates may also communicate independent investment recommendations, market color or trading ideas and/or publish or express independent research views in respect of such assets, securities or instruments and may at any time hold, or recommend to clients that they should acquire, long and/or short positions in such assets, securities and instruments.

A prospectus in electronic format may be made available on the web sites maintained by one or more underwriters, or selling group members, if any, participating in the offering. The underwriters may agree to allocate a number of shares to underwriters and selling group members for sale to their online brokerage account holders. Internet distributions will be allocated by the representatives to underwriters and selling group members that may make Internet distributions on the same basis as other allocations.

Sales of shares made outside of the United States may be made by affiliates of the underwriters. Other than in the United States, no action has been taken by us or the underwriters that would permit a public offering of the securities offered by this prospectus in any jurisdiction where action for that purpose is required. The securities offered by this prospectus may not be offered or sold, directly or indirectly, nor may this prospectus or any other offering material or advertisements in connection with the offer and sale of any such securities be distributed or published in any jurisdiction, except under circumstances that will result in compliance with the applicable rules and regulations of that jurisdiction. Persons into whose possession this prospectus comes are advised to inform themselves about and to observe any restrictions relating to the offering and the distribution of this prospectus. This prospectus does not constitute an offer to sell or a solicitation of an offer to buy any securities offered by this prospectus in any jurisdiction in which such an offer or a solicitation is unlawful.

Selling restrictions

European Economic Area

In relation to each Member State of the European Economic Area which has implemented the Prospectus Directive (each, a "Relevant Member State") an offer to the public of our Class A common shares may not be made in that Relevant Member State, except that an offer to the public in that Relevant Member State of our Class A common shares may be made at any time under the following exemptions under the Prospectus Directive:

- To any legal entity which is a qualified investor as defined in the Prospectus Directive;
- To fewer than 150 natural or legal persons (other than qualified investors as defined in the Prospectus Directive), subject to obtaining the prior consent of the J.P. Morgan Securities LLC and Goldman Sachs & Co. LLC for any such offer; or

- In any other circumstances falling within Article 3(2) of the Prospectus Directive;

provided that no such offer of our common shares shall result in a requirement for the publication by us or any Brazilian placement agent of a prospectus pursuant to Article 3 of the Prospectus Directive.

For the purposes of this provision, the expression an "offer to public" in relation to our Class A common shares in any Relevant Member State means the communication in any form and by any means of sufficient information on the terms of the offer and our Class A common shares to be offered so as to enable an investor to decide to purchase our Class A common shares, as the same may be varied in that Member State by any measure implementing the Prospectus Directive in that Member State, the expression "Prospectus Directive" means Directive 2003/71/EC (as amended), including by Directive 2010/73/EU and includes any relevant implementing measure in the Relevant Member State.

This European Economic Area selling restriction is in addition to any other selling restrictions set out below.

United Kingdom

In the United Kingdom, this prospectus is only addressed to and directed as qualified investors who are (i) investment professionals falling within Article 19(5) of the Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 (the Order); or (ii) high net worth entities and other persons to whom it may lawfully be communicated, falling within Article 49(2)(a) to (d) of the Order (all such persons together being referred to as "relevant persons"). Any investment or investment activity to which this prospectus relates is available only to relevant persons and will only be engaged with relevant persons. Any person who is not a relevant person should not act or relay on this prospectus or any of its contents.

Canada

The securities may be sold in Canada only to purchasers purchasing, or deemed to be purchasing, as principal that are accredited investors, as defined in National Instrument 45-106 Prospectus Exemptions or subsection 73.3(1) of the Securities Act (Ontario), and are permitted clients, as defined in National Instrument 31-103 Registration Requirements, Exemptions, and Ongoing Registrant Obligations. Any resale of the securities must be made in accordance with an exemption from, or in a transaction not subject to, the prospectus requirements of applicable securities laws.

Securities legislation in certain provinces or territories of Canada may provide a purchaser with remedies for rescission or damages if this prospectus (including any amendment thereto) contains a misrepresentation, provided that the remedies for rescission or damages are exercised by the purchaser within the time limit prescribed by the securities legislation of the purchaser's province or territory. The purchaser should refer to any applicable provisions of the securities legislation of the purchaser's province or territory of these rights or consult with a legal advisor.

Pursuant to section 3A.3 of National Instrument 33-105 Underwriting Conflicts (NI 33-105), the underwriters are not required to comply with the disclosure requirements of NI 33-105 regarding underwriter conflicts of interest in connection with this offering.

Hong Kong

The shares may not be offered or sold in Hong Kong by means of any document other than (i) in circumstances which do not constitute an offer to the public within the meaning of the Companies (Winding Up and Miscellaneous Provisions) Ordinance (Cap. 32 of the Laws of Hong Kong) ("Companies (Winding Up and Miscellaneous Provisions) Ordinance") or which do not constitute an invitation to the public within the

meaning of the Securities and Futures Ordinance (Cap. 571 of the Laws of Hong Kong) ("Securities and Futures Ordinance"), or (ii) to "professional investors" as defined in the Securities and Futures Ordinance and any rules made thereunder, or (iii) in other circumstances which do not result in the document being a "prospectus" as defined in the Companies (Winding Up and Miscellaneous Provisions) Ordinance, and no advertisement, invitation or document relating to the shares may be issued or may be in the possession of any person for the purpose of issue (in each case whether in Hong Kong or elsewhere), which is directed at, or the contents of which are likely to be accessed or read by, the public in Hong Kong (except if permitted to do so under the securities laws of Hong Kong) other than with respect to shares which are or are intended to be disposed of only to persons outside Hong Kong or only to "professional investors" in Hong Kong as defined in the Securities and Futures Ordinance and any rules made thereunder.

Singapore

This prospectus has not been registered as a prospectus with the Monetary Authority of Singapore. Accordingly, this prospectus and any other document or material in connection with the offer or sale, or invitation for subscription or purchase, of the shares may not be circulated or distributed, nor may the shares be offered or sold, or be made the subject of an invitation for subscription or purchase, whether directly or indirectly, to persons in Singapore other than (i) to an institutional investor (as defined under Section 4A of the Securities and Futures Act, Chapter 289 of Singapore (the "SFA")) under Section 274 of the SFA, (ii) to a relevant person (as defined in Section 275(2) of the SFA) pursuant to Section 275(1) of the SFA, or any person pursuant to Section 275(1A) of the SFA, and in accordance with the conditions specified in Section 275 of the SFA or (iii) otherwise pursuant to, and in accordance with the conditions of, any other applicable provision of the SFA, in each case subject to conditions set forth in the SFA.

Where the shares are subscribed or purchased under Section 275 of the SFA by a relevant person which is a corporation (which is not an accredited investor (as defined in Section 4A of the SFA)) the sole business of which is to hold investments and the entire share capital of which is owned by one or more individuals, each of whom is an accredited investor, the securities (as defined in Section 239(1) of the SFA) of that corporation shall not be transferable for 6 months after that corporation has acquired the shares under Section 275 of the SFA except: (1) to an institutional investor under Section 274 of the SFA or to a relevant person (as defined in Section 275(2) of the SFA), (2) where such transfer arises from an offer in that corporation's securities pursuant to Section 275(1A) of the SFA, (3) where no consideration is or will be given for the transfer, (4) where the transfer is by operation of law, (5) as specified in Section 276(7) of the SFA, or (6) as specified in Regulation 32 of the Securities and Futures (Offers of Investments) (Shares and Debentures) Regulations 2005 of Singapore ("Regulation 32").

Where the shares are subscribed or purchased under Section 275 of the SFA by a relevant person which is a trust (where the trustee is not an accredited investor (as defined in Section 4A of the SFA)) whose sole purpose is to hold investments and each beneficiary of the trust is an accredited investor, the beneficiaries' rights and interest (howsoever described) in that trust shall not be transferable for 6 months after that trust has acquired the shares under Section 275 of the SFA except: (1) to an institutional investor under Section 274 of the SFA or to a relevant person (as defined in Section 275(2) of the SFA), (2) where such transfer arises from an offer that is made on terms that such rights or interest are acquired at a consideration of not less than $200,000 (or its equivalent in a foreign currency) for each transaction (whether such amount is to be paid for in cash or by exchange of securities or other assets), (3) where no consideration is or will be given for the transfer, (4) where the transfer is by operation of law, (5) as specified in Section 276(7) of the SFA, or (6) as specified in Regulation 32.

Japan

The securities have not been and will not be registered under the Financial Instruments and Exchange Act of Japan (Act No. 25 of 1948, as amended), or the FIEA. The securities may not be offered or sold, directly or indirectly, in Japan or to or for the benefit of any resident of Japan (including any person resident in Japan or any corporation or other entity organized under the laws of Japan) or to others for reoffering or resale, directly or indirectly, in Japan or to or for the benefit of any resident of Japan, except pursuant to an exemption from the registration requirements of the FIEA and otherwise in compliance with any relevant laws and regulations of Japan.

Legal matters

The validity of the Class A common shares offered hereby will be passed upon for us by Conyers Dill & Pearman Limited. Certain legal matters as to U.S. law in connection with this offering will be passed upon for us by Latham & Watkins LLP. Certain legal matters as to U.S. law in connection with this offering will be passed upon for the underwriters by Ropes & Gray LLP.

Experts

The financial statements as of December 31, 2016 and 2017 and for the years then ended included in this prospectus have been so included in reliance on the report of PricewaterhouseCoopers LLP, an independent registered public accounting firm, given on the authority of said firm as experts in auditing and accounting.

Exchange controls

The permission of the Bermuda Monetary Authority is required, pursuant to the provisions of the Exchange Control Act 1972 and related regulations, for all issuances and transfers of shares (which includes our Class A common shares) of Bermuda companies to or from a non-resident of Bermuda for exchange control purposes, other than in cases where the Bermuda Monetary Authority has granted a general permission. The Bermuda Monetary Authority has, pursuant to its statement of June 1, 2005, given its general permission under the Exchange Control Act 1972 (and related regulations) for the issue and free transferability of our Class A common shares to and between non-residents of Bermuda for exchange control purposes, provided that the Class A common shares remain listed on an appointed stock exchange, which includes The Nasdaq Global Select Market. Approvals or permissions given by the Bermuda Monetary Authority do not constitute a guarantee by the Bermuda Monetary Authority as to our performance or our creditworthiness. Accordingly, in giving such consent or permissions, the Bermuda Monetary Authority shall not be liable for the financial soundness, performance or default of our business or for the correctness of any opinions or statements expressed herein. Certain issues and transfers of Class A common shares involving persons deemed resident in Bermuda for exchange control purposes require the specific consent of the Bermuda Monetary Authority.

Enforcement of civil liabilities under United States federal securities laws

We are organized pursuant to the laws of Bermuda. In addition, it is anticipated that some or all of our directors and officers will reside outside the United States, and all or a substantial portion of our assets and their assets are or may be located in jurisdictions outside the United States. As a result, it may be difficult for you to effect service of process within the United States upon those persons or us or to recover against them or us on judgments of United States courts, including judgments predicated upon civil liability provisions of the United States federal securities laws.

We have been advised that there is no treaty in force between the United States and Bermuda providing for the reciprocal recognition and enforcement of judgments in civil and commercial matters. As a result, whether a U.S. judgment would be enforceable in Bermuda against us or our directors and officers depends on whether the U.S. court that entered the judgment is recognized by the Bermuda court as having jurisdiction over us or our directors and officers, as determined by reference to Bermuda conflict of law rules. A judgment debt from a U.S. court that is final and for a sum certain based on U.S. federal securities laws will not be enforceable in Bermuda unless the judgment debtor had submitted to the jurisdiction of the U.S. court, and the issue of submission and jurisdiction is a matter of Bermuda (not U.S.) law.

In addition, and irrespective of jurisdictional issues, the Bermuda courts will not enforce a U.S. federal securities law that is either penal or contrary to Bermuda public policy. We have been advised that an action brought pursuant to a public or penal law, the purpose of which is the enforcement of a sanction, power or right at the instance of the state in its sovereign capacity, will not be entertained by a Bermuda court. Certain remedies available under the laws of U.S. jurisdictions, including certain remedies under U.S. federal securities laws, would not be available under Bermuda law or enforceable in a Bermuda court, as they would be contrary to Bermuda public policy. Further, no claim may be brought in Bermuda against us or our directors and officers in the first instance for violation of U.S. federal securities laws because these laws have no extraterritorial jurisdiction under Bermuda law and do not have force of law in Bermuda. A Bermuda court may, however, impose civil liability on us or our directors and officers if the facts alleged in a complaint constitute or give rise to a cause of action under Bermuda law.

Where you can find more information

We have filed with the SEC a registration statement on Form S-1 under the Securities Act with respect to the Class A common shares offered hereby. This prospectus, which constitutes a part of the registration statement, does not contain all of the information set forth in the registration statement or the exhibits and schedules filed therewith. For further information about us and the Class A common shares offered hereby, we refer you to the registration statement and the exhibits and schedules filed thereto. Statements contained in this prospectus regarding the contents of any contract or any other document that is filed as an exhibit to the registration statement are not necessarily complete, and each such statement is qualified in all respects by reference to the full text of such contract or other document filed as an exhibit to the registration statement. We are subject to the information requirements of the Securities Exchange Act of 1934, as amended, and, in accordance therewith, file periodic reports, proxy statements, and other information with the SEC These documents may be accessed through the SEC's electronic data gathering, analysis and retrieval system, or EDGAR, via electronic means, including the SEC's home page on the Internet (*www.sec.gov*).

Our website is located at *www.kiniksa.com*. The reference to our website address does not constitute incorporation by reference of the information contained at or available through our website, and you should not consider it to be a part of this prospectus.

Index to consolidated financial statements

Annual consolidated financial statements as of December 31, 2016 and 2017

Interim consolidated financial statements as of September 30, 2017 and 2018 (unaudited)

Report of independent registered public accounting firm

To the Board of Directors and Shareholders of
Kiniksa Pharmaceuticals, Ltd.

Opinion on the financial statements

We have audited the accompanying consolidated balance sheets of Kiniksa Pharmaceuticals, Ltd. and its subsidiary (the "Company") as of December 31, 2017 and 2016, and the related consolidated statements of operations and comprehensive loss, of convertible preferred shares and shareholders' deficit and of cash flows for the years then ended, including the related notes (collectively referred to as the "consolidated financial statements"). In our opinion, the consolidated financial statements present fairly, in all material respects, the financial position of the Company as of December 31, 2017 and 2016, and the results of its operations and its cash flows for the years then ended in conformity with accounting principles generally accepted in the United States of America.

Basis for opinion

These consolidated financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on the Company's consolidated financial statements based on our audits. We are a public accounting firm registered with the Public Company Accounting Oversight Board (United States) ("PCAOB") and are required to be independent with respect to the Company in accordance with the U.S. federal securities laws and the applicable rules and regulations of the Securities and Exchange Commission and the PCAOB.

We conducted our audits of these consolidated financial statements in accordance with the standards of the PCAOB. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the consolidated financial statements are free of material misstatement, whether due to error or fraud.

Our audits included performing procedures to assess the risks of material misstatement of the consolidated financial statements, whether due to error or fraud, and performing procedures that respond to those risks. Such procedures included examining, on a test basis, evidence regarding the amounts and disclosures in the consolidated financial statements. Our audits also included evaluating the accounting principles used and significant estimates made by management, as well as evaluating the overall presentation of the consolidated financial statements. We believe that our audits provide a reasonable basis for our opinion.

/s/ PricewaterhouseCoopers LLP
Boston, Massachusetts
February 27, 2018, except for the effects of the reverse share split
discussed in Note 14 to the consolidated financial statements, as to
which the date is May 14, 2018

We have served as the Company's auditor since 2016.

Kiniksa Pharmaceuticals, Ltd.
Consolidated balance sheets
(In thousands, except share and per share amounts)

	December 31, 2016	December 31, 2017
Assets		
Current assets:		
Cash and cash equivalents	$ 55,970	$ 45,555
Restricted cash	105	105
Prepaid expenses and other current assets	259	1,444
Total current assets	56,334	47,104
Property and equipment, net	84	125
Restricted cash	—	—
Deferred offering costs	8	25
Deferred tax assets	41	238
Total assets	$ 56,467	$ 47,492
Liabilities, Convertible Preferred Shares and Shareholders' Equity (Deficit)		
Current liabilities:		
Accounts payable	$ 212	$ 1,218
Accrued expenses	2,090	6,212
Accrued milestone	—	10,000
Total current liabilities	2,302	17,430
Deferred rent	—	—
Total liabilities	2,302	17,430
Commitments and contingencies (Note 12)		
Convertible preferred shares (Series A, B and C), $0.000273235 par value; 17,128,120 shares and 22,885,492 shares designated, issued and outstanding as of December 31, 2016 and 2017, respectively; aggregate liquidation preference of $80,000 and $120,000 as of December 31, 2016 and 2017	79,897	119,770
Shareholders' equity (deficit):		
Class A common shares, par value of $0.000273235 per share; 4,491,921 shares and 5,507,938 shares designated as of December 31, 2016 and 2017, respectively; 719,976 shares issued and outstanding as of December 31, 2016 and 2017	—	—
Class B common shares, par value of $0.000273235 per share; 3,568,353 shares designated, issued and outstanding as of December 31, 2016 and 2017	1	1
Class A1 common shares, $0.000273235 par value; no shares designated, issued or outstanding as of December 31, 2016 and 2017	—	—
Class B1 common shares, $0.000273235 par value; no shares designated, issued or outstanding as of December 31, 2016 and 2017	—	—
Additional paid-in capital	392	1,289
Accumulated deficit	(26,125)	(90,998)
Total shareholders' equity (deficit)	(25,732)	(89,708)
Total liabilities, convertible preferred shares and shareholders' equity (deficit)	$ 56,467	$ 47,492

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

Kiniksa Pharmaceuticals, Ltd.
Consolidated statements of operations and comprehensive loss
(In thousands, except share and per share amounts)

	Year ended December 31,	
	2016	**2017**
Operating expenses:		
Research and development	$ 17,439	$ 56,357
General and administrative	6,563	9,043
Total operating expenses	24,002	65,400
Loss from operations	(24,002)	(65,400)
Interest income	65	529
Loss before provision for income taxes	(23,937)	(64,871)
Provision for income taxes	(36)	(2)
Net loss and comprehensive loss	$ (23,973)	$ (64,873)
Net loss per share attributable to common shareholders—basic and diluted	$ (91.61)	$ (35.85)
Weighted average common shares outstanding—basic and diluted	261,695	1,809,751

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

Kiniksa Pharmaceuticals, Ltd.
Consolidated statements of convertible preferred shares and shareholders' deficit
(In thousands, except share amounts)

	Convertible preferred shares (Series A, B and C)		Common shares (Class A and B)		Additional paid-In capital	Accumulated deficit	Total shareholders' deficit
	Shares	Amount	Shares	Amount			
Balances at December 31, 2015	8,028,809	$ 37,398	4,282,020	$ 1	$ 14	$ (2,152)	$ (2,137)
Issuance of Series A convertible preferred shares, net of issuance costs of $1	9,099,311	42,499	—	—	—	—	—
Exercise of options	—	—	6,309	—	10	—	10
Share-based compensation expense	—	—	—	—	368	—	368
Net loss	—	—	—	—	—	(23,973)	(23,973)
Balances at December 31, 2016	17,128,120	79,897	4,288,329	1	392	(26,125)	(25,732)
Issuance of Series B convertible preferred shares, net of issuance costs of $127	5,757,372	39,873	—	—	—	—	—
Share-based compensation expense	—	—	—	—	897	—	897
Net loss	—	—	—	—	—	(64,873)	(64,873)
Balances at December 31, 2017	22,885,492	119,770	4,288,329	1	1,289	(90,998)	(89,708)

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

Kiniksa Pharmaceuticals, Ltd.
Consolidated statements of cash flows
(In thousands)

	Year ended December 31,	
	2016	2017
Cash flows from operating activities:		
Net loss	$ (23,973)	$ (64,873)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	22	28
Share-based compensation expense	368	897
Deferred income taxes	(46)	(197)
Changes in operating assets and liabilities:		
Prepaid expenses and other current assets	(219)	(1,185)
Accounts payable	119	1,006
Accrued expenses	1,862	4,105
Accrued milestone	—	10,000
Net cash used in operating activities	(21,867)	(50,219)
Cash flows from investing activities:		
Purchases of property and equipment	(3)	(69)
Net cash used in investing activities	(3)	(69)
Cash flows from financing activities:		
Proceeds from issuance of Series A convertible preferred shares, net of issuance costs	42,499	—
Proceeds from issuance of Series B convertible preferred shares, net of issuance costs	—	39,873
Proceeds from exercise of options	10	—
Net cash provided by financing activities	42,509	39,873
Net increase (decrease) in cash and cash equivalents and restricted cash	20,639	(10,415)
Cash and cash equivalents and restricted cash at beginning of year	35,436	56,075
Cash and cash equivalents and restricted cash at end of year	$ 56,075	$ 45,660
Supplemental information:		
Cash paid for income taxes	$ 115	$ 290
Supplemental disclosure of non-cash investing and financing activities:		
Deferred offering costs included in accrued expenses and accounts payable	$ 8	$ 25

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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements

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

1. Nature of the business and basis of presentation

Kiniksa Pharmaceuticals, Ltd. (the "Company") is a clinical-stage biopharmaceutical company focused on 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, currently focused on autoinflammatory and autoimmune conditions.

The Company is subject to risks and uncertainties common to early-stage companies in the biotechnology industry. There can be no assurance that the Company's research and development will be successfully completed, that adequate protection for the Company's technology will be obtained, that any products developed will obtain necessary government regulatory approval or that any products, if approved, will be commercially viable. The Company operates in an environment of rapid technological innovation and substantial competition from pharmaceutical and biotechnological companies. In addition, the Company is dependent upon the services of its employees, consultants and service providers. Even if the Company's product development efforts are successful, it is uncertain when, if ever, the Company will realize significant revenue from product sales.

Through December 31, 2017, the Company has funded its operations primarily with proceeds from the sale of convertible preferred shares. The Company has incurred recurring losses since its inception, including net losses of $23,973 and $64,873 for the years ended December 31, 2016 and 2017. In addition, as of December 31, 2017, the Company had an accumulated deficit of $90,998. The Company expects to continue to generate operating losses for the foreseeable future. As of February 27, 2018, the issuance date of the annual consolidated financial statements for the years ended December 31, 2016 and 2017, the Company expected that its cash and cash equivalents of $45,555 as of December 31, 2017, together with the $200,000 of gross proceeds received from the Company's sale of Series C convertible preferred shares in February 2018 (see Note 6), would be sufficient to fund its operating expenses and capital expenditure requirements for at least twelve months from the issuance date of the annual consolidated financial statements. The future viability of the Company beyond that point is dependent on its ability to raise additional capital to finance its operations.

The Company is seeking to complete an initial public offering ("IPO") of its common shares. Upon the closing of a qualified public offering, on specified terms, the Company's outstanding convertible preferred shares will automatically convert into common shares (see Note 6). In the event the Company does not complete an IPO, the Company expects to seek additional funding through private equity financings, debt financings, or other capital sources, which may include collaborations with other companies, government funding arrangements or other strategic transactions. The Company may not be able to obtain financing on acceptable terms, or at all. The terms of any financing may adversely affect the holdings or the rights of the Company's shareholders.

If the Company is unable to obtain funding, the Company will be forced to delay, reduce or eliminate some or all of its research and development programs, product portfolio expansion or commercialization efforts, which could adversely affect its business prospects, or the Company may be unable to continue operations. Although management continues to pursue these plans, there is no assurance that the Company will be successful in obtaining sufficient funding on terms acceptable to the Company to fund continuing operations, if at all.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

2. Summary of significant accounting policies

Principles of consolidation

The accompanying consolidated financial statements have been prepared in accordance with accounting principles generally accepted in the United States of America ("GAAP") and include the accounts of the Company and its wholly owned U.S. subsidiary, Kiniksa Pharmaceuticals Corp. ("Kiniksa US"), after elimination of all significant intercompany accounts and transactions.

In assessing the consolidation requirement for variable interest entities ("VIEs"), the Company focuses on identifying whether it has both the power to direct the activities that most significantly impact the VIE's economic performance and the obligation to absorb losses or the right to receive benefits from the VIE. In the event that the Company is the primary beneficiary of a VIE, the assets, liabilities, and results of operations of the VIE would be included in the Company's consolidated financial statements. At December 31, 2016 and 2017 and during the years then ended, the Company was not the primary beneficiary of a VIE.

Use of estimates

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

Cash and cash equivalents

The Company classifies deposits in banks, money market funds and cash invested temporarily in various instruments with maturities of three months or less at the time of purchase as cash and cash equivalents. At December 31, 2016 and 2017, cash and cash equivalents consisted principally of U.S. Treasury notes, amounts held in money market accounts and cash on deposit at commercial banks.

Concentrations of credit risk

Financial instruments that potentially subject the Company to concentrations of credit risk consist principally of cash and cash equivalents. At December 31, 2016 and 2017, all of the Company's cash and cash equivalents were held at two financial institutions. The Company generally maintains balances in various operating accounts at financial institutions that management believes to be of high credit quality, in amounts that may exceed federally insured limits. The Company has not experienced any losses related to its cash and cash equivalents and does not believe that it is subject to unusual credit risk beyond the normal credit risk associated with commercial banking relationships.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

Restricted cash

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

Property and equipment

Property and equipment are recorded at cost and depreciated over the estimated useful lives of the related assets using the straight-line method. Maintenance and repairs are charged to expense as incurred. When assets are retired or otherwise disposed of, the cost of these assets and related accumulated depreciation or amortization are eliminated from the consolidated balance sheet and any resulting gains or losses are included in the consolidated statement of operations and comprehensive loss in the period of disposal. The expected useful lives of the respective assets are as follows:

	Estimated useful life
Computer hardware and software	3 - 5 years
Vehicles	5 years
Laboratory and facility equipment	5 years
Furniture and fixtures	5 - 7 years
Leasehold improvements	Shorter of estimated useful life or lease term

Impairment of long-lived assets

Long-lived assets consist of property and equipment. Long-lived assets to be held and used are tested for recoverability whenever events or changes in business circumstances indicate that the carrying amount of the assets may not be fully recoverable. Factors that the Company considers in deciding when to perform an impairment review include significant underperformance of the business in relation to expectations, significant negative industry or economic trends and significant changes or planned changes in the use of the assets. If an impairment review is performed to evaluate a long-lived asset group for recoverability, the Company compares forecasts of undiscounted cash flows expected to result from the use and eventual disposition of the long-lived asset group to its carrying value. An impairment loss would be recognized when estimated undiscounted future cash flows expected to result from the use of an asset group are less than its carrying amount. The impairment loss would be based on the excess of the carrying value of the impaired asset group over its fair value, determined based on discounted cash flows. To date, the Company has not recorded any impairment losses on long-lived assets.

Deferred offering costs

The Company capitalizes certain legal, professional accounting and other third-party fees that are directly associated with in-process preferred share or common equity financings as deferred offering costs until such financings are consummated. After consummation of the equity financing, these costs are recorded as a reduction to the carrying value of convertible preferred shares or in shareholders' equity (deficit) as a reduction of additional paid-in capital generated as a result of the offering. Should an in-process equity

financing be abandoned, the deferred offering costs will be expensed immediately as a charge to operating expenses in the consolidated statements of operations and comprehensive loss. The Company recorded deferred offering costs related to the sale of convertible preferred shares of $8 and $25 as of December 31, 2016 and 2017, respectively.

Fair value measurements

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

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

The Company's restricted cash, which is held in a money market fund, is carried at fair value, determined based on Level 1 inputs in the fair value hierarchy described above (see Note 3). The Company's cash equivalents, consisting of money market accounts and U.S. Treasury notes, are carried at fair value, determined based on Level 1 and 2 inputs in the fair value hierarchy described above (see Note 3). The carrying values of the Company's prepaid expenses and other current assets, accounts payable and accrued expenses approximate their fair values due to the short-term nature of these assets and liabilities.

Segment information

The Company manages its operations as a single operating segment for the purposes of assessing performance and making operating decisions. The Company's singular focus is on developing and delivering therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need.

Research and development costs

Research and development costs are expensed as incurred. Research and development expenses consist of costs incurred to discover, research and develop drug candidates, including personnel expenses, share-based compensation expense, allocated facility-related and depreciation expenses, third-party license fees and external costs of outside vendors engaged to conduct clinical development activities and clinical trials as well as to manufacture clinical trial materials. Non-refundable prepayments for goods or services that will be used or rendered for future research and development activities are recorded as prepaid expenses.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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.

Patent costs

The Company charges patent-related costs in connection with filing and prosecuting patent applications to operations as incurred as their realization is uncertain. These costs are classified as general and administrative expenses.

Share-based compensation

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

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

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

The fair value of each restricted share award is estimated on the date of grant based on the fair value of the Company's Class A or Class B common shares on that same date. The fair value of each option grant is estimated on the date of grant using the Black-Scholes option-pricing model, which requires inputs based on certain subjective assumptions, including the expected share price volatility, the expected term of the award, the risk-free interest rate, and expected dividends (see Note 8). The Company historically has been a private company and lacks company-specific historical and implied volatility information for its shares.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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

Comprehensive loss

Comprehensive loss includes net loss as well as other changes in shareholders' equity (deficit) that result from transactions and economic events other than those with shareholders. There was no difference between net loss and comprehensive loss for each of the periods presented in the accompanying consolidated financial statements.

Income taxes

The Company accounts for income taxes using the asset and liability method, which requires the recognition of deferred tax assets and liabilities for the expected future tax consequences of events that have been recognized in the consolidated financial statements or in the Company's tax returns. Deferred tax assets and liabilities are determined based on the difference between the financial statement and tax basis of assets and liabilities using enacted tax rates in effect for the year in which the differences are expected to reverse. Changes in deferred tax assets and liabilities are recorded in the provision for income taxes. The Company assesses the likelihood that its deferred tax assets will be recovered from future taxable income and, to the extent it believes, based upon the weight of available evidence, that it is more likely than not that all or a portion of the deferred tax assets will not be realized, a valuation allowance is established through a charge to income tax expense. Potential for recovery of deferred tax assets is evaluated by estimating the future taxable profits expected and considering prudent and feasible tax planning strategies.

The Company accounts for uncertainty in income taxes recognized in the consolidated financial statements by applying a two-step process to determine the amount of tax benefit to be recognized. First, the tax position must be evaluated to determine the likelihood that it will be sustained upon external examination by the taxing authorities. If the tax position is deemed more-likely-than-not to be sustained, the tax position is then assessed to determine the amount of benefit to recognize in the consolidated financial statements. The amount of the benefit that may be recognized is the largest amount that has a greater than 50% likelihood of being realized upon ultimate settlement. The provision for income taxes includes the effects of any resulting tax reserves, or unrecognized tax benefits, that are considered appropriate as well as the related net interest and penalties. To date, the Company has not taken any uncertain tax positions or recorded any reserves, interest or penalties.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

Net income (loss) per share

The Company follows the two-class method when computing net income (loss) per share as the Company has issued shares that meet the definition of participating securities. The two-class method determines net income (loss) per share for each class of common and participating securities according to dividends declared or accumulated and participation rights in undistributed earnings. The two-class method requires income available to common shareholders for the period to be allocated between common and participating securities based upon their respective rights to receive dividends as if all income for the period had been distributed.

Basic net income (loss) per share attributable to common shareholders is computed by dividing the net income (loss) attributable to common shareholders by the weighted average number of common shares outstanding for the period. Diluted net income (loss) attributable to common shareholders is computed by adjusting net income (loss) attributable to common shareholders to reallocate undistributed earnings based on the potential impact of dilutive securities. Diluted net income (loss) per share attributable to common shareholders is computed by dividing the diluted net income (loss) attributable to common shareholders by the weighted average number of common shares outstanding for the period, including potential dilutive common shares. For purpose of this calculation, outstanding options, unvested restricted common shares and convertible preferred shares are considered potential dilutive common shares.

The Company's convertible preferred shares contractually entitle the holders of such shares to participate in dividends but do not contractually require the holders of such shares to participate in losses of the Company. Accordingly, in periods in which the Company reports a net loss attributable to common shareholders, such losses are not allocated to such participating securities. In periods in which the Company reports a net loss attributable to common shareholders, diluted net loss per share attributable to common shareholders is the same as basic net loss per share attributable to common shareholders, since dilutive common shares are not assumed to have been issued if their effect is anti-dilutive. The Company reported a net loss attributable to common shareholders for the years ended December 31, 2016 and 2017.

Recently adopted accounting pronouncements

In May 2017, the Financial Accounting Standards Board ("FASB") issued Accounting Standards Update ("ASU") No. 2017-09, Compensation—Stock Compensation (Topic 718): Scope of Modification Accounting ("ASU 2017-09"), which clarifies when to account for a change to the terms or conditions of a share-based payment award as a modification. Under the new guidance, modification accounting is required only if the fair value, the vesting conditions or the classification of the award (as equity or liability) changes as a result of the change in terms or conditions. ASU 2017-09 is effective for annual periods beginning after December 15, 2017, including interim periods within those fiscal years. Early adoption is permitted. The Company adopted ASU 2017-09 as of the required effective date of January 1, 2018. The adoption of ASU 2017-09 will have an impact on the modification of stock-based awards, if any, after the date of adoption. The adoption of ASU 2017-09 did not have an impact on the Company's financial position, results of operations or cash flows.

In January 2017, the FASB issued ASU No. 2017-01, *Business Combinations (Topic 805) Clarifying the Definition of a Business* ("ASU 2017-01"). ASU 2017-01 clarifies the definition of a business by adding

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

guidance to assist entities in evaluating whether transactions should be accounted for as acquisitions of assets or businesses. The definition of a business affects many areas of accounting including acquisitions, disposals, goodwill and consolidation. The ASU is effective for fiscal years beginning after December 15, 2018. Early adoption is permitted. The Company adopted this standard effective as of January 1, 2016 and applied it to its license and asset purchase agreements during the years ended December 31, 2016 and 2017 (see Note 9).

In November 2016, the FASB issued ASU No. 2016-18, *Statement of Cash Flows (Topic 230): Restricted Cash* ("ASU 2016-18"), which requires that amounts generally described as restricted cash and restricted cash equivalents be included with cash and cash equivalents when reconciling the beginning-of-period and end-of-period total amounts shown on the statement of cash flows. ASU 2016-18 is effective for fiscal years beginning after December 15, 2017 and interim periods within those fiscal years and should be applied using a retrospective transition method to each period presented. Early adoption is permitted. The Company elected to early adopt ASU 2016-18 effective as of January 1, 2017 and has reflected the adoption retrospectively to all periods presented in its consolidated financial statements. As a result, the Company's consolidated statements of cash flows include restricted cash with cash and cash equivalents when reconciling the beginning-of-period and end-of-period total amounts shown on the such statements.

In August 2016, the FASB issued ASU No. 2016-15, Statement of Cash Flows (Topic 230): Classification of Certain Cash Receipts and Cash Payments ("ASU 2016-15"). This guidance addresses diversity in practice in how certain cash receipts and cash payments are presented in the statement of cash flows. The standard is effective for fiscal years beginning after December 15, 2017, including interim periods in those fiscal years, and early adoption is permitted. The adoption of ASU 2016-15 is required to be applied retrospectively. The Company adopted ASU 2017-09 as of the required effective date of January 1, 2018, and the adoption did not have an impact on the Company's consolidated statement of cash flows.

In March 2016, the FASB issued ASU No. 2016-09, *Compensation—Stock Compensation (Topic 718): Improvements to Employee Share-Based Payment Accounting* ("ASU 2016-09"). ASU 2016-09 addresses several aspects of the accounting for share-based payment transactions, including the income tax consequences, classification of awards as either equity or liabilities, an option to recognize gross share compensation expense with actual forfeitures recognized as they occur, and classification on the statement of cash flows. Certain of these changes are required to be applied retrospectively, while other changes are required to be applied prospectively. The Company adopted ASU 2016-09 effective as of January 1, 2016 and elected prospectively to account for forfeitures as they occur rather than apply an estimated forfeiture rate to share-based compensation expense. The adoption of ASU 2016-09 did not have a material impact on the Company's financial position, results of operations or cash flows.

In November 2015, the FASB issued ASU No. 2015-17*, Income Taxes (Topic 740): Balance Sheet Classification of Deferred Taxes* ("ASU 2015-17"). ASU 2015-17 requires deferred tax liabilities and assets to be classified as non-current in the consolidated balance sheet. The amendment may be applied either prospectively to all deferred tax liabilities and assets or retrospectively to all periods presented. The Company adopted ASU 2015-17 effective as of January 1, 2016 and has reflected the adoption retrospectively to all periods presented in its consolidated financial statements. The adoption of

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

ASU 2015-17 did not have a material impact on the Company's financial position, results of operations or cash flows.

In August 2014, the FASB issued ASU No. 2014-15, *Presentation of Financial Statements—Going Concern (Subtopic 205-40): Disclosure of Uncertainties about an Entity's Ability to Continue as a Going Concern* ("ASU 2014-15"). The amendments in this update explicitly require a company's management to assess if there is substantial doubt about an entity's ability to continue as a going concern within one year of the date of issuance of the entity's financial statements and to provide related footnote disclosures in certain circumstances. The Company adopted ASU 2014-15 effective as of January 1, 2016. This guidance relates to footnote disclosure only and its adoption had no impact on the Company's financial position, results of operations or cash flows.

In May 2014, the FASB issued ASU No. 2014-09*, Revenue from Contracts with Customers (Topic 606)* ("ASU 2014-09"), which supersedes existing revenue recognition guidance under GAAP. The standard's core principle is that a company will recognize revenue when it transfers promised goods or services to customers in an amount that reflects the consideration to which the company expects to be entitled in exchange for those goods or services. The standard defines a five-step process to achieve this principle, and will require companies to use more judgment and make more estimates than under the current guidance. The Company expects that these judgments and estimates will include identifying performance obligations in the customer contract, estimating the amount of variable consideration to include in the transaction price and allocating the transaction price to each separate performance obligation. ASU 2014-09 also requires additional disclosure about the nature, amount, timing and uncertainty of revenue and cash flows arising from customer contracts. In August 2015, the FASB issued ASU No. 2015-14, *Revenue from Contracts with Customers (Topic 606): Deferral of the Effective Date*, which delays the effective date of ASU 2014-09 such that the standard is effective for public entities for annual periods beginning after December 15, 2017 and for interim periods within those fiscal years. The FASB subsequently issued amendments to ASU 2014-09 that have the same effective date and transition date. The Company adopted ASU 2014-09 as of the required effective date of January 1, 2018 and the adoption did not have an impact on the Company's consolidated financial statements as the Company does not currently have any revenue-generating arrangements.

Recently issued accounting pronouncements

In July 2017, the FASB issued ASU No. 2017-11, *Earnings Per Share (Topic 260), Distinguishing Liabilities from Equity (Topic 480), Derivatives and Hedging (Topic 815) I. Accounting for Certain Financial Instruments with Down Round Features II. Replacement of the Indefinite Deferral for Mandatorily Redeemable Financial Instruments of Certain Nonpublic Entities and Certain Mandatorily Redeemable Noncontrolling Interests with a Scope Exception ("ASU 2017-11").* Part I applies to entities that issue financial instruments such as warrants, convertible debt or convertible preferred stock that contain down-round features. Part II replaces the indefinite deferral for certain mandatorily redeemable noncontrolling interests and mandatorily redeemable financial instruments of nonpublic entities contained within ASC Topic 480 with a scope exception and does not impact the accounting for these mandatorily redeemable instruments. ASU 2017-11 is required to be adopted for annual periods beginning after

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

December 15, 2018, including interim periods within those fiscal years. The Company is currently evaluating the impact that the adoption of ASU 2017-11 will have on its consolidated financial statements.

In February 2016, the FASB issued ASU No. 2016-02, *Leases (Topic 842)* ("ASU 2016-02"), which sets out the principles for the recognition, measurement, presentation and disclosure of leases for both parties to a contract (i.e., lessees and lessors). The new standard requires lessees to apply a dual approach, classifying leases as either finance or operating leases based on the principle of whether or not the lease is effectively a financed purchase by the lessee. This classification will determine whether lease expense is recognized based on an effective interest method or on a straight-line basis over the term of the lease, respectively. A lessee is also required to record a right-of-use asset and a lease liability for all leases with a term of greater than 12 months regardless of their classification. Leases with a term of 12 months or less will be accounted for similar to existing guidance for operating leases today. The guidance is effective for annual periods beginning after December 15, 2018 and for interim periods within those fiscal years, and early adoption is permitted. The Company is currently evaluating the impact that the adoption of ASU 2016-02 will have on its consolidated financial statements.

3. Fair value of financial assets and liabilities

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

	Fair value measurements as of December 31, 2016 using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash—money market funds	$ 105	$ —	$ —	$ 105
Cash equivalents—money market funds	550	—	—	550
Cash equivalents—U.S. Treasury notes	—	52,504	—	52,504
	$ 655	$ 52,504	$ —	$ 53,159

	Fair value measurements as of December 31, 2017 using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash—money market funds	$ 105	$ —	$ —	$ 105
Cash equivalents—money market funds	5,487	—	—	5,487
Cash equivalents—U.S. Treasury notes	—	14,995	—	14,995
	$ 5,592	$ 14,995	$ —	$ 20,587

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

During the years ended December 31, 2016 and 2017, there were no transfers between Level 1, Level 2 and Level 3.

The money market funds were valued using quoted prices in active markets, which represent a Level 1 measurement in the fair value hierarchy. The Company's cash equivalents as of December 31, 2016 and 2017 also consisted of U.S. Treasury notes, which are not traded on a daily basis and, therefore, represent a Level 2 measurement in the fair value hierarchy at each period end.

4. Property and equipment, net

Property and equipment, net consisted of the following:

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

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

5. Accrued expenses

Accrued expenses consisted of the following:

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

6. Convertible preferred shares

As of December 31, 2017, the Company's bye-laws, as amended and restated (the "Amended Bye-Laws"), 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 have been further designated as Series A convertible preferred shares (the "Series A preferred shares") and 5,757,372 shares have been further designated as Series B convertible preferred shares (the "Series B preferred shares"). The holders of preferred shares

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have liquidation rights in the event of a deemed liquidation that, in certain situations, are not solely within the control of the Company. Therefore, the Series A and Series B preferred shares (collectively, the "Preferred Shares") are classified outside of shareholders' equity (deficit).

In October 2015, the Company issued and sold 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 convertible 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.

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

	December 31, 2016				
	Preferred shares designated	Preferred shares issued and outstanding	Carrying value	Liquidation preference	Common shares issuable upon conversion
Series A preferred shares	17,128,120	17,128,120	$ 79,897	$ 80,000	17,128,120

	December 31, 2017				
	Preferred shares designated	Preferred shares issued and outstanding	Carrying value	Liquidation preference	Common shares issuable upon conversion
Series A preferred shares	17,128,120	17,128,120	$ 79,897	$ 80,000	17,128,120
Series B preferred shares	5,757,372	5,757,372	39,873	40,000	5,757,372
	22,885,492	22,885,492	$ 119,770	$ 120,000	22,885,492

The holders of the Preferred Shares have the following rights and preferences:

Voting

The holders of Preferred Shares are entitled to vote, together with the holders of common shares, on all matters submitted to shareholders for a vote. The holders of Series A preferred shares are entitled to the number of votes per Series A preferred share equal to the number of whole Class B common shares into which the Series A preferred shares are convertible on the record date determining shareholders entitled to participate in such vote (which is ten votes for each Class B common share). The holders of Series B preferred shares are entitled to the number of votes per Series B preferred share equal to the number of

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whole Class A common shares into which the Series B preferred shares are convertible on the record date determining shareholders entitled to participate in such vote (which is one vote for each Class A common share). Except as provided by law or by the other provisions of the Amended Bye-Laws, holders of Preferred Shares vote together with the holders of common shares as a single class.

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

Conversion

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

The Series A Original Issue Price and Series A Conversion Price were equal to $4.6707 as of December 31, 2016 and 2017. The Series B Original Issue Price and Series B Conversion Price were equal to $6.9475 as of December 31, 2017. Such Series A and Series B Original Issue Prices and Series A and Series B Conversion Prices, and the rate at which each series of preferred shares may be converted into common shares, are subject to adjustment from time to time to reflect future share dividends, splits, combinations, recapitalizations and similar events. The Series A and Series B Conversion Prices are also subject to adjustments based on weighted-average anti-dilution provisions set forth in the Amended Bye-Laws in the event that additional securities are issued at a purchase price less than the Series A Conversion Price and/or the Series B Conversion Price then in effect. As of December 31, 2016 and 2017, each Series A preferred share was convertible into one Class B common share. As of December 31, 2017, each Series B preferred share was convertible into one Class A common share.

Upon either (a) the closing of the sale of Class A common shares or Class B common shares to the public at a price of at least $14.0120 per share (subject to appropriate adjustment in the event of any share dividend, share split, combination or other similar recapitalization with respect to the applicable class of common shares) in an initial public offering resulting in at least $100,000 of gross proceeds to the Company (a "Qualified IPO") or (b) the date and time, or the occurrence of an event, specified by vote or written consent of the holders of a majority of the outstanding Preferred Shares, voting together as a single class on an as-if-converted to Class A common shares basis, all outstanding Series A preferred shares shall automatically be converted, in such manner as is permitted pursuant to Bermuda law, into Class B common shares at the then effective conversion rate, and all outstanding Series B preferred shares

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shall automatically be converted, in such manner as is permitted pursuant to Bermuda law, into Class A common shares at the then effective conversion rate. In the event of a mandatory conversion of preferred shares as a result of a Qualified IPO, (i) holders of Series A preferred shares may elect to receive Class B1 common shares in lieu of Class B common shares and (ii) holders of Series B preferred shares may elect to receive Class A1 common shares in lieu of Class A common shares.

Dividends

The holders of the Preferred Shares are entitled to receive noncumulative dividends when and if declared by Company's board of directors. The Company may not declare, pay or set aside any dividends on any other class or series of shares of the Company, other than dividends on common shares payable in common shares, unless the holders of the Preferred Share first receive, or simultaneously receive, a dividend on each outstanding Preferred Share equal to (A) in the case of a dividend on any class of common shares or any class or series that is convertible into common shares, that dividend per Preferred Share as would equal the product of (1) the dividend payable on each share of such class or series determined, if applicable, as if all shares of such class or series had been converted into common shares and (2) the number of common shares issuable upon conversion of a share the applicable series of Preferred Shares, or (B) in the case of a dividend on any class or series that is not convertible into common shares, at a rate per Preferred Share determined by (1) dividing the amount of the dividend payable on each share of such class or series of shares by the original issue price of such class or series (subject to appropriate adjustment in the event of any bonus share, share dividend, share split, combination of or other similar recapitalization with respect to such class or series) and (2) multiplying such fraction by an amount equal to the applicable Series A or Series B Original Issue Price. Through December 31, 2016 and 2017, no cash dividends have been declared or paid.

Liquidation preference

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

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

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Unless a majority of the holders of the then outstanding Preferred Shares, on an as-if-converted to Class A common shares basis, elect otherwise, a deemed liquidation event shall include a merger or consolidation (other than one in which shareholders of the Company own a majority by voting power of the outstanding shares of the surviving or acquiring company or corporation) or a sale, lease, transfer, exclusive license or other disposition of all or substantially all of the assets of the Company.

Redemption

The Amended Bye-Laws do not provide redemption rights to the holders of Preferred Shares.

7. Common shares

As of December 31, 2016, and 2017, the Amended Bye-Laws authorized the Company to issue 43,918,239 total shares with a par value of $0.000273235, of which 4,491,921 and 5,507,938 shares have been designated as Class A common shares as of December 31, 2016 and 2017, respectively, and 3,568,353 shares have been designated as Class B common shares as of December 31, 2016 and 2017. The remaining 18,729,845 and 11,956,456 shares that were not designated as common shares or Preferred Shares as of December 31, 2016 and 2017, respectively, may be designated to any class at any time in the future by the Company's board of directors. No Class A1 common shares or Class B1 common shares were designated as of December 31, 2016 and 2017. The rights of the holders of the Company's Class A common shares, Class B common shares, Class A1 common shares and Class B1 common shares are identical, except with respect to voting and conversion, as described below. The voting, dividend and liquidation rights of the holders of the Company's common shares are subject to and qualified by the rights, powers and preferences of the holders of the Preferred Shares as set forth above.

Voting

Each Class A common share entitles the holder to one vote on all matters submitted to the shareholders for a vote. Each Class B common share entitles the holder to ten votes on all matters submitted to the shareholders for a vote. Holders of Class A1 common shares or Class B1 common shares have no voting rights. The holders of Class A and Class B common shares, voting together as a single class, are entitled to elect one director of the Company.

Dividends

Common shareholders are entitled to receive dividends, as may be declared by the board of directors. These dividends are subject to the preferential dividend rights of the holders of the Company's Preferred Shares. Through December 31, 2016 and 2017, no cash dividends have been declared or paid.

Conversion

Each Class B common share shall automatically convert into one Class A common share upon certain transfers of such shares by the holder thereof (subject to certain exceptions). Each Class B common share shall be convertible, at the holder's election and at any time into one Class A common share or one Class B1 common share. Each Class A1 common share is convertible into one Class A common share at the

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holder's election. Each Class B1 common share is convertible into one Class A common share or one Class B common share at the holder's election.

There are no conversion rights associated with the Company's Class A common shares.

8. Share-based compensation

2015 equity incentive plan

The Company's 2015 Equity Incentive Plan, as amended (the "2015 Plan"), provides for the Company to grant qualified incentive options, nonqualified options, share grants and other share-based awards to employees and non-employees to purchase the Company's Class A common shares.

The total number of common shares that may be issued under the 2015 Plan was 3,778,249 and 4,794,266 shares as of December 31, 2016 and 2017, respectively, of which 2,188,249 shares remained available for future grant as of December 31, 2016 and 1,664,893 shares remained available for future grant as of December 31, 2017.

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

Shares that are expired, terminated, surrendered or canceled under the 2015 Plan without having been fully exercised will be available for future awards.

During the years ended December 31, 2016 and 2017, the Company granted options to purchase 316,866 and 1,545,045 Class A common shares, respectively, to employees and directors. The Company recorded share-based compensation expense for options granted to employees and directors of $354 and $876 during the years ended December 31, 2016 and 2017, respectively.

During the years ended December 31, 2016 and 2017, the Company granted options to purchase 12,807 and 1,829 Class A common shares, respectively, to non-employees. The Company recorded share-based compensation expense for options granted to non-employees of $14 and $21 during the years ended December 31, 2016 and 2017, respectively.

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

Option valuation

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

	Year ended December 31,	
	2016	2017
Risk-free interest rate	1.45%	1.99%
Expected term (in years)	6.25	6.25
Expected volatility	70.75%	74.18%
Expected dividend yield	0%	0%

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

	Year ended December 31,	
	2016	2017
Risk-free interest rate	1.94%	2.49%
Expected term (in years)	10.00	10.00
Expected volatility	65.85%	78.28%
Expected dividend yield	0%	0%

Options

Through December 31, 2017, all options granted by the Company under the 2015 Plan were for the purchase of Class A common shares. The following table summarizes option activity under the 2015 Plan for the year ended December 31, 2017:

	Number of shares	Weighted average exercise price	Weighted average remaining contractual term	Aggregate intrinsic value
			(in years)	
Outstanding as of December 31, 2016	1,583,691	$ 1.64	9.10	$ 356
Granted	1,546,874	3.90		
Exercised	—	—		
Forfeited	(7,501)	3.75		
Outstanding as of December 31, 2017	3,123,064	$ 2.75	8.82	$ 6,010
Options exercisable as of December 31, 2017	843,454	$ 1.62	8.07	$ 2,580
Options unvested as of December 31, 2017	2,279,610	$ 3.17	9.09	$ 3,431

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The aggregate intrinsic value of options is calculated as the difference between the exercise price of the options and the fair value of the Company's common shares for those options that had exercise prices lower than the fair value of the Company's common shares.

During the year ended December 31, 2016, an option holder exercised 6,309 options for Class A common shares with an intrinsic value of $2 for total cash proceeds of $10. There were no options exercised during the year ended December 31, 2017.

The weighted-average grant-date fair value per share of options granted during the years ended December 31, 2016 and 2017 was $1.16 and $2.57, respectively.

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

Restricted shares

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

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

	Class A		Class B	
	Number of shares	Weighted average fair value at issuance	Number of shares	Weighted average fair value at issuance
Unvested restricted shares outstanding as of December 31, 2016	490,646	$ 0.000273235	2,527,583	$ 0.000273235
Granted	—	—	—	—
Vested	(178,417)	0.000273235	(892,088)	0.000273235
Unvested restricted shares outstanding as of December 31, 2017	312,229	$ 0.000273235	1,635,495	$ 0.000273235

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

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

Share-based compensation

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

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

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

9. License and acquisition agreements

Biogen asset purchase agreement

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to KPL-716 and other antibodies covered by certain patent rights, including patents and other intellectual property rights, clinical data, know-how, and clinical drug supply. In addition, Biogen granted to the Company a non-exclusive, sublicensable, worldwide 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. During the year ended December 31, 2017, the Company made a milestone payment of $4,000 associated with the achievement of a specified clinical milestone event. Additionally, the Company could be obligated to make up to an aggregate of up to $150,000 of payments upon the achievement of specified annual net sales milestones and to pay tiered royalties 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

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sublicense fee of $150 and is obligated to pay insignificant annual maintenance fees as well as clinical and regulatory milestone payments of up to an aggregate of $1,575. During the year ended December 31, 2017 the Company paid $75 upon the achievement of certain milestones in connection with the retained contracts.

The Biogen Agreement will terminate upon the expiration of all payment obligations with respect to the last product in all countries in the territory. The Company has the right to terminate the agreement with 90 days' prior written notice. Both parties may terminate by mutual written consent or in the event of material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches).

During the years ended December 31, 2016 and 2017, the Company recorded research and development expense in connection with the Biogen Agreement of $12,100 and $4,169, respectively.

Novo Nordisk license agreement

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

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

Under the Novo Nordisk Agreement, the Company is also required to make a payment of $150 upon completion of the technology transfer by Novo Nordisk. In addition, the Company is obligated to make milestone payments upon the achievement of specified clinical, regulatory and initial sales milestones and upon the achievement of annual net sales thresholds, including a payment of $1,000 upon the earlier to occur of a specified regulatory milestone and January 2020, unless the Novo Nordisk Agreement is earlier terminated by either party. As of December 31, 2017, the Company determined that the payment related to the milestone was not probable and, therefore, no amount was recorded in the Company's consolidated statement of operations and comprehensive loss during the year ended December 31, 2017. The Company has also agreed to pay royalties on annual net sales of products licensed under the agreement.

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

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

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licensed patent rights. The Company may also terminate the agreement for any reason upon prior written notice to Novo Nordisk.

During the year ended December 31, 2017, the Company recorded research and development expense of $1,500, in connection with the Novo Nordisk Agreement.

Primatope stock purchase option agreement

In September 2017, the Company entered into a stock purchase option agreement (the "Primatope Agreement") with Primatope Therapeutics, Inc. ("Primatope"), pursuant to which the Company has been granted a license to certain intellectual property rights controlled by Primatope to research, develop, and manufacture the pre-clinical antibody, KPL-404.

The agreement provides the Company with an exclusive call option to purchase 100% of the capital stock of Primatope. Upon execution of the agreement, the Company made $500 in upfront payments for the initial option period through April 2018 (the "Initial Option Period"). The Primatope Agreement allows up to three extensions of the Initial Option Period through January 2019 (including the initial option period, the "Option Period") for total extension payments of up to $800. During the Option Period, the Company may conduct research and pre-clinical work to assess the viability of the asset.

If the call option is exercised, the Company will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10,000 as well as potential milestone payments of up to $10,000. The upfront payment and the milestone payments may be paid in a combination of cash and issuance of the Company's Class A common shares.

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

Either party may terminate the Primatope Agreement for uncured material breach of the agreement by the other party or by mutual written consent.

During the year ended December 31, 2017, the Company recorded research and development expense of $500 in connection with the Primatope Agreement.

Regeneron license agreement

In September 2017, the Company entered into a license agreement (the "Regeneron Agreement") with Regeneron Pharmaceuticals, Inc. ("Regeneron"), pursuant to which the Company has been granted an exclusive, sublicensable license under certain intellectual property rights controlled by Regeneron to develop and commercialize 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

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business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

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

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

The parties also entered into a clinical supply agreement under which Regeneron agreed to manufacture the developed product during the clinical phase. During the year ended December 31, 2017, the Company recognized research and development expense of $208 related to the purchase of drug materials under this agreement. As of December 31, 2017, the Company has non-cancelable purchase commitments under the clinical supply agreement (see Note 12).

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

During the year ended December 31, 2017, the Company recorded research and development expense of $5,208, in connection with the agreements with Regeneron.

MedImmune license agreement

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

In exchange for these rights, the Company made an upfront payment of $8,000. The Company accounted for the acquisition of technology as an asset acquisition because it did not meet the definition of a

business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use. In addition, the Company is obligated to make clinical, regulatory and initial sales milestone payments of up to $72,500 in aggregate for the first two indications, including a milestone payment of $10,000 upon the earlier to occur of a specified regulatory milestone and December 31, 2018, unless the MedImmune Agreement is earlier terminated by either party. As of December 31, 2017, the Company determined that the payment related to this milestone was probable and, therefore, recognized research and development expense and an accrued milestone of $10,000 during the year ended December 31, 2017. In addition, the Company is obligated to make clinical and regulatory milestone payments of up to $15,000 in the aggregate for each subsequent indication. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties 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 year ended December 31, 2017, the Company recorded research and development expense of $18,000 in connection with the MedImmune Agreement.

10. Income taxes

As a company incorporated in Bermuda, the Company is principally subject to taxation in Bermuda. Under the current laws of Bermuda, tax on a company's income is assessed at a zero percent tax rate. As a result, the Company has not recorded any income tax benefits from its losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards will be available to the Company for those losses.

In August 2015, the Company entered into agreements with its wholly owned subsidiary, Kiniksa US, under which Kiniksa US provides management and research and development services to the Company for which the Company pays costs plus a service fee. Kiniksa US is subject to tax for federal and state tax purposes.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

On December 22, 2017, the United States enacted new tax reform ("Tax Cuts and Jobs Act"). The Tax Cuts and Jobs Act contains provisions with separate effective dates but is generally effective for taxable years beginning after December 31, 2017. Beginning with the year ending December 31, 2018, the corporate statutory rates on U.S. earnings will be reduced from a top marginal rate of 35% to a flat rate of 21%. The impact of the future rate reduction resulted in a provision for income taxes of $69 for the year ended December 31, 2017 relating to the revaluation of the Company's net deferred tax assets.

Income (loss) before provision for income taxes consisted of the following:

	Year ended December 31,	
	2016	2017
Bermuda	$ (24,254)	$ (65,391)
Foreign (U.S.)	317	520
	$ (23,937)	$ (64,871)

The components of the Company's income tax provision for the years ended December 31, 2016 and 2017 are as follows:

	Year ended December 31,	
	2016	2017
Current income tax provision:		
Bermuda	$ —	$ —
U.S. federal	78	184
U.S. state	4	15
Total current income tax provision	82	199
Deferred income tax provision (benefit):		
Bermuda	—	—
U.S. federal	(26)	(87)
U.S. state	(20)	(110)
Total deferred income tax provision (benefit)	(46)	(197)
Total provision for income taxes	$ 36	$ 2

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

A reconciliation of the Bermuda statutory income tax rate of 0% to the Company's effective income tax rate is as follows:

	Year ended December 31,	
	2016	2017
Bermuda statutory income tax rate	—%	—%
Foreign (U.S.) tax rate differential	(0.5)	(0.4)
Research and development tax credits	0.5	0.5
2017 Tax Cuts and Jobs Act	—	(0.1)
Effective income tax rate	—%	—%

Net deferred tax assets consisted of the following:

	December 31,	
	2016	2017
Research and development tax credit carryforwards	$ 18	$ 90
Depreciation and amortization	(4)	(14)
Accrued expenses and other	37	189
Total deferred tax assets	51	265
Valuation allowance	(10)	(27)
Net deferred tax assets	$ 41	$ 238

As of December 31, 2017, the Company had state research and development tax credit carryforwards of approximately $113, available to reduce future tax liabilities, which begin to expire in 2031 through 2032.

As required by ASC 740, the Company has evaluated the positive and negative evidence bearing upon its ability to realize the deferred tax assets. In order to utilize state research and development tax credits, the Company will need taxable income in the jurisdiction of where the credit was generated. The Company currently has no taxable income in certain state jurisdictions and thus management has determined that it is more likely than not that the Company will not recognize the benefits of state research and development tax credits generated in those jurisdictions, and as a result, a valuation allowance of $10 and $27 has been established at December 31, 2016 and 2017, respectively. The remaining deferred tax assets will be fully utilized in the United States based on future income generated under the cost-plus arrangement in place.

Utilization of the state research and development tax credits may be subject to substantial annual limitation under Section 382 of the Internal Revenue Code of 1986 due to ownership changes that could occur in the future. These ownership changes may limit the amount of carryforwards that can be utilized annually to offset future taxable income. In general, an ownership change, as defined by Section 382, results from transactions increasing the ownership of certain shareholders or public groups in the stock of a corporation by more than 50% over a three-year period.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

Changes in the valuation allowance for deferred tax assets during the years ended December 31, 2016 and 2017 were due primarily to an increase in state research and development tax credits and were as follows:

	Year ended December 31,	
	2016	2017
Valuation allowance at beginning of year	$ (1)	$ (10)
Increases recorded to income tax provision	(9)	(17)
Valuation allowance at end of year	$ (10)	$ (27)

The Company has not recorded any amounts for unrecognized tax benefits as of December 31, 2016 or 2017. The Company's policy is to record interest and penalties related to income taxes as part of its income tax provision. As of December 31, 2016 and 2017, the Company had no accrued interest or penalties related to uncertain tax positions and no amounts had been recognized in the Company's consolidated statements of operations and comprehensive loss.

The Company files income tax returns in the United States and certain state jurisdictions. Kiniksa US's federal and state income tax returns are subject to tax examinations for the tax years ended December 31, 2013 and subsequent years. To the extent the Company has tax attribute carryforwards, the tax years in which the attribute was generated may still be adjusted upon examination by the Internal Revenue Service and state tax authorities to the extent utilized in a future period. There are currently no income tax examinations pending.

11. Net loss per share

The rights, including the liquidation and dividend rights, of the holders of Class A and Class B common shares are identical, except with respect to voting rights. As the liquidation and dividend rights are identical, losses are allocated on a proportionate basis and the resulting net loss per share attributed to common shareholders will, therefore, be the same for both Class A and Class B common shares on an individual or combined basis.

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

	Year ended December 31,	
	2016	2017
Numerator:		
Net loss attributable to common shareholders	$ (23,973)	$ (64,873)
Denominator:		
Weighted average common shares outstanding—basic and diluted	261,695	1,809,751
Net loss per share attributable to common shareholders—basic and diluted	$ (91.61)	$ (35.85)

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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

	Year ended December 31,	
	2016	2017
Options to purchase common shares	1,583,691	3,123,064
Unvested restricted shares	3,018,229	1,947,724
Convertible preferred shares (as converted to common shares)	17,128,120	22,885,492
	21,730,040	27,956,280

12. Commitments and contingencies

Lease agreements

On July 24, 2015, Kiniksa US entered into an operating lease in Wellesley Hills, Massachusetts for office space that comprises the headquarters for Kiniksa US. In March 2016, effective August 1, 2016, Kiniksa US entered into an expansion and extension on its lease, which expanded its leased space to a total of 10,800 square feet. On March 31, 2017, Kiniksa US renewed this lease and extended the lease term to August 2018. Monthly lease payments, inclusive of base rent and ancillary charges, total $27. As of December 31, 2017 future minimum lease payments under non-cancelable operating lease commitments, which are all due during the year ending December 31, 2018, totaled $270.

The Company recognizes rent expense on a straight-line basis over the respective lease period. The Company recorded rent expense of $286 and $402 during the years ended December 31, 2016 and 2017, respectively.

License agreements

The Company has entered into license agreements with various parties under which it is obligated to make contingent and non-contingent payments (see Note 9).

Manufacturing commitments

During the year ended December 31, 2017, the Company entered into agreements with several contract manufacturing organizations to provide pre-clinical and clinical trial materials. As of December 31, 2017,

the Company had non-cancelable purchase commitments under these agreements totaling $7,766, which are all due during the year ending December 31, 2018.

Indemnification agreements

In the ordinary course of business, the Company may provide indemnification of varying scope and terms to vendors, lessors, business partners and other parties with respect to certain matters including, but not limited to, losses arising out of breach of such agreements or from intellectual property infringement claims made by third parties. In addition, the Company has entered into indemnification agreements with members of its board of directors and executive officers that will require the Company, among other things, to indemnify them against certain liabilities that may arise by reason of their status or service as directors or officers. The maximum potential amount of future payments the Company could be required to make under these indemnification agreements is, in many cases, unlimited. To date, the Company has not incurred any material costs as a result of such indemnifications. The Company does not believe that the outcome of any claims under indemnification arrangements will have a material effect on its financial position, results of operations or cash flows, and it has not accrued any liabilities related to such obligations in its consolidated financial statements as of December 31, 2016 or 2017.

Legal proceedings

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

13. Benefit plans

The Company has established a defined-contribution savings plan under Section 401(k) of the Internal Revenue Code. This plan covers substantially all employees who meet minimum age and service requirements and allows participants to defer a portion of their annual compensation on a pre-tax basis. The Company provides matching contributions of 100% of the first 3% of each participant's salary contributed, plus 50% for each of the next 2% contributed. Employees are immediately and fully vested in their own contributions and the Company's match. During the years ended December 31, 2016 and 2017, the Company contributed $143 and $264, respectively, to the plan.

14. Subsequent events

For its consolidated financial statements as of December 31, 2017 and for the year then ended, the Company evaluated subsequent events through February 27, 2018, the date on which those financial statements were issued, and, with respect to the reverse share split described below, through May 14, 2018.

Sale of Series C preferred shares

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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

The rights and preferences of the Series C preferred shares are substantially similar to the Company's Preferred Shares. Each Series C preferred share shall be convertible, at the option of the holder, at any time and from time to time, and without the payment of additional consideration by the holder thereof, in such manner as is permitted by Bermuda law, into such number of fully paid and non-assessable Class A common shares as is determined by dividing the Series C original issue price by the Series C conversion price, each initially equal to $15.6438, in effect at the time of conversion. In the event of a mandatory conversion of Series C preferred shares as a result of a Qualified IPO, holders of Series C preferred shares may elect to receive Class AI common shares in lieu of Class A common shares.

In February 2018, in connection with the Company's sale of Series C preferred shares, the Company amended and restated its Amended Bye-Laws to increase the total number of authorized shares of all classes of capital stock to 44,746,463 shares, consisting of 35,670,093 preferred shares, 5,507,938 Class A common shares and 3,568,353 Class B common shares.

Reverse share 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.

Kiniksa Pharmaceuticals, Ltd.
Consolidated balance sheets
(In thousands, except share and per share amounts)
(Unaudited)

	September 30, 2018	December 31, 2017
Assets		
Current assets:		
Cash and cash equivalents	$ 69,663	$ 45,555
Restricted cash	—	105
Short-term investments	268,200	—
Prepaid expenses and other current assets	3,665	1,444
Total current assets	341,528	47,104
Property and equipment, net	2,360	125
Restricted cash	210	—
Deferred offering costs	—	25
Deferred tax assets	1,173	238
Advanced clinical payments	1,830	—
Total assets	$ 347,101	$ 47,492
Liabilities, Convertible Preferred Shares and Shareholders' Equity (Deficit)		
Current liabilities:		
Accounts payable	$ 4,668	$ 1,218
Accrued expenses	13,353	6,212
Accrued milestone	10,000	10,000
Total current liabilities	28,021	17,430
Deferred rent	167	—
Total liabilities	28,188	17,430
Commitments and contingencies (Note 11)		
Convertible preferred shares (Series A, B and C), $0.000273235 par value; 0 shares and 22,885,492 shares issued and outstanding as of September 30, 2018 and December 31, 2017, respectively; aggregate liquidation preference of $0 and $120,000 as of September 30, 2018 and December 31, 2017, respectively;	—	119,770
Shareholders' equity (deficit):		
Class A common shares, par value of $0.000273235 per share; 15,772,257 shares and 719,976 shares issued and outstanding as of September 30, 2018 and December 31, 2017; respectively	4	—
Class B common shares, par value of $0.000273235 per share; 4,638,855 shares and 3,568,353 shares issued and outstanding as of September 30, 2018 and December 31, 2017, respectively	1	1
Class A1 common shares, $0.000273235 par value; 12,995,954 shares and 0 shares issued and outstanding as of September 30, 2018 and December 31, 2017, respectively	4	—
Class B1 common shares, $0.000273235 par value; 16,057,618 shares and 0 shares issued and outstanding as of September 30, 2018 and December 31, 2017, respectively	4	—
Additional paid-in capital	470,600	1,289
Accumulated other comprehensive loss	(55)	—
Accumulated deficit	(151,645)	(90,998)
Total shareholders' equity (deficit)	318,913	(89,708)
Total liabilities, convertible preferred shares and shareholders' equity (deficit)	$ 347,101	$ 47,492

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

Kiniksa Pharmaceuticals, Ltd.
Consolidated statements of operations and comprehensive loss
(In thousands, except share and per share amounts)
(Unaudited)

	Three months ended September 30,		Nine months ended September 30,	
	2018	2017	2018	2017
Operating expenses:				
Research and development	$ 20,644	$ 14,008	$ 50,475	$ 26,426
General and administrative	5,515	2,241	13,550	6,263
Total operating expenses	26,159	16,249	64,025	32,689
Loss from operations	(26,159)	(16,249)	(64,025)	(32,689)
Interest income	1,622	169	2,992	396
Loss before provision for income taxes	(24,537)	(16,080)	(61,033)	(32,293)
Benefit for income taxes	131	51	386	121
Net loss and comprehensive loss	$ (24,406)	$ (16,029)	$ (60,647)	$ (32,172)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.51)	$ (8.25)	$ (2.62)	$ (19.21)
Weighted average common shares outstanding—basic and diluted	48,183,424	1,942,106	23,174,841	1,675,133

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

Kiniksa Pharmaceuticals, LTD.
Consolidated statements of convertible preferred shares and shareholders' equity (deficit)
(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 OCL	Accumulated deficit	Total shareholders' equity (deficit)
	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	—	—	—	—	—	—
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	155,532	—	—	155,536
Exercise of options	—	—	22,060	—	76	—	—	76
Share-based compensation expense	—	—	—	—	3,119	—	—	3,119
Unrealized loss on short-term investments	—	—	—	—	—	(55)	—	(55)
Net loss	—	—	—	—	—	—	(60,647)	(60,647)
Balances at September 30, 2018	—	$ —	49,464,684	$13	$470,600	$(55)	$(151,645)	$ 318,913

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

Kiniksa Pharmaceuticals, Ltd.
Consolidated statements of cash flows
(In thousands)
(Unaudited)

	Nine months ended September 30,	
	2018	**2017**
Cash flows from operating activities:		
Net loss	$ (60,647)	$ (32,172)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	32	20
Share-based compensation expense	3,119	550
Loss on disposal of property and equipment	66	—
Non-cash rent expense	258	—
Accretion of discounts on short-term investments	(670)	—
Deferred income taxes	(935)	(168)
Changes in operating assets and liabilities:		
Prepaid expenses and other assets	(4,027)	(1,406)
Accounts payable	2,981	6,013
Accrued expenses	6,449	323
Net cash used in operating activities	(53,374)	(26,840)
Cash flows from investing activities:		
Purchases of property and equipment	(1,274)	(65)
Purchases of short-term investments	(292,584)	—
Proceeds from the maturities of short-term investments	25,000	—
Net cash used in investing activities	(268,858)	(65)
Cash flows from financing activities:		
Proceeds from issuance of Series B convertible preferred shares, net of issuance costs	—	39,873
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	159,193	—
Payments of deferred offering costs	(3,646)	—
Proceeds from exercise of options	76	—
Net cash provided by financing activities	346,445	39,873
Net increase (decrease) in cash and cash equivalents and restricted cash	24,213	12,968
Cash and cash equivalents and restricted cash at beginning of period	45,660	56,075
Cash and cash equivalents and restricted cash at end of period	$ 69,873	$ 69,043
Supplemental information:		
Cash paid for income taxes	$ 345	$ —
Supplemental disclosure of non-cash investing and financing activities:		
Deferred offering costs included in accrued expenses and accounts payable	$ 11	$ —
Property and equipment included in accrued expenses and accounts payable	1,058	—

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, currently focused on autoinflammatory and autoimmune conditions.

The Company is subject to risks and uncertainties common to early-stage companies in the biotechnology industry. There can be no assurance that the Company's research and development will be successfully completed, that adequate protection for the Company's technology will be obtained, that any products developed will obtain necessary government regulatory approval or that any products, if approved, will be commercially viable. The Company 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 biotechnological companies. In addition, the Company is dependent upon the services of its employees, consultants and service providers. Even if the Company's product development efforts are successful, it is uncertain when, if ever, the Company will realize significant revenue from product sales.

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 subsidiary, Kiniksa Pharmaceuticals Corp. ("Kiniksa US"), after elimination of all significant intercompany accounts and transactions.

In assessing the consolidation requirement for variable interest entities ("VIEs"), the Company focuses on identifying whether it has both the power to direct the activities that most significantly impact the VIE's economic performance and the obligation to absorb losses or the right to receive benefits from the VIE. In the event that the Company is the primary beneficiary of a VIE, the assets, liabilities, and results of operations of the VIE would be included in the Company's consolidated financial statements. At December 31, 2017 and during the year then ended and at September 30, 2018 and during the three and nine 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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

periodically reviewed in light of changes in circumstances, facts and experience. Changes in estimates are recorded in the period in which they become known. Actual results could differ from those estimates.

Unaudited interim consolidated financial information

The accompanying unaudited consolidated financial statements have been prepared in accordance with GAAP for interim financial information. The accompanying unaudited consolidated financial statements do not include all of the information and footnotes required by GAAP for complete consolidated financial statements. The accompanying year-end consolidated balance sheet was derived from audited financial statements, but does not include all disclosures required by GAAP. The unaudited interim consolidated financial statements have been prepared on the same basis as the audited annual consolidated financial statements and, in the opinion of management, reflect all adjustments, which include only normal recurring adjustments, necessary for the fair statement of the Company's financial position as of September 30, 2018 and the results of its operations for the three and nine months ended September 30, 2018 and 2017 and its cash flows for the nine months ended September 30, 2018 and 2017. The results for the three and nine months ended September 30, 2018 are not necessarily indicative of results to be expected for the year ending December 31, 2018, any other interim periods or any future year or period.

Reverse share 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 $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 of their over-allotment option to purchase additional shares at $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 ("Amended & Restated Bye-Laws").

Liquidity

In accordance with Accounting Standards Update ("ASU") 2014-15, Disclosure of Uncertainties about an Entity's Ability to Continue as a Going Concern (Subtopic 205-40), the Company has evaluated whether

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

there are conditions and events, considered in the aggregate, that raise substantial doubt about the Company's ability to continue as a going concern within one year after the date the consolidated financial statements are issued. As of September 30, 2018, the Company had an accumulated deficit of $151,645. During the nine months ended September 30, 2018, the Company incurred a net loss of $60,647 and used $53,374 of net cash in operating activities. The Company expects to continue to generate operating losses for the foreseeable future. As of September 30, 2018, the Company had cash, cash equivalents and short-term investments of $337,863. Based on its current operating plan, the Company expects this amount will be sufficient to fund its operating expenses and capital expenditure requirements for at least twelve months from the issuance date of these consolidated financial statements. The future viability of the Company beyond that point is dependent on its ability to raise additional capital to finance its operations. 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 September 30, 2018 and December 31, 2017, 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 and are reported at fair value with unrealized gains and losses included as a component of shareholders' equity (deficit). Realized gains and losses, if any, on short-term investments are included in interest income (expense), net.

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.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

Concentrations of credit risk

Financial instruments that potentially subject the Company to concentrations of credit risk consist principally of cash, cash equivalents and short-term investments. At September 30, 2018 and December 31, 2017, 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

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

In conjunction with the Company's lease agreement entered into in March 2018 (see Note 11), the Company maintains a letter of credit for the benefit of the landlord. As of September 30, 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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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(see Note 3). The carrying values of the Company's prepaid expenses and other current assets, accounts payable and accrued expenses approximate their fair values due to the short-term nature of these assets and liabilities.

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 pre-clinical 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.

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Notes to consolidated financial statements (Continued)

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

The fair value of each restricted share award is estimated on the date of grant based on the fair value of the Company's Class A or Class B common shares on that same date. The fair value of each option grant is estimated using the Black-Scholes option-pricing model, which requires inputs based on certain subjective assumptions, including the expected share price volatility, the expected term of the award, the risk-free interest rate, and expected dividends (see Note 8). Prior to May 2018, the Company was a private company and, accordingly, lacks company-specific historical and implied volatility information for its shares. Therefore, it estimates its expected share price volatility based on the historical volatility of publicly traded peer companies and expects to continue to do so until such time as it has adequate historical data regarding the volatility of its own traded share price. The expected term of the Company's options has been determined utilizing the "simplified" method for awards that qualify as "plain-vanilla" options. The expected term of options granted to non-employees is equal to the contractual term of the option award. The risk-free interest rate is determined by reference to the U.S. Treasury yield curve in effect at the time of grant of the award for time periods approximately equal to the expected term of the award. Expected dividend yield is based on the fact that the Company has never paid cash dividends on common shares and does not expect to pay any cash dividends in the foreseeable future.

Net income (loss) per share

The Company follows the two-class method when computing net income (loss) per share as the Company has issued shares that meet the definition of participating securities. The two-class method determines net income (loss) per share for each class of common and participating securities according to dividends declared or accumulated and participation rights in undistributed earnings. The two-class method requires income available to common shareholders for the period to be allocated between common and participating securities based upon their respective rights to receive dividends as if all income for the period had been distributed.

Basic net income (loss) per share attributable to common shareholders is computed by dividing the net income (loss) attributable to common shareholders by the weighted average number of common shares outstanding for the period. Diluted net income (loss) attributable to common shareholders is computed by adjusting net income (loss) attributable to common shareholders to reallocate undistributed earnings based on the potential impact of dilutive securities. Diluted net income (loss) per share attributable to common shareholders is computed by dividing the diluted net income (loss) attributable to common shareholders by the weighted average number of common shares outstanding for the period, including potential dilutive common shares. For purpose of this calculation, outstanding options, unvested restricted common shares and convertible preferred shares are considered potential dilutive common shares.

Prior to the closing of its 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

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Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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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 nine months ended September 30, 2018 and 2017.

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, however the Company will revise the three and six month periods ended June 30, 2018 the next time they are presented. This revision did not impact the loss per share for the three months ended September 30, 2018, however it resulted in an increase of $0.09 loss per share for the nine months ended September 30, 2018 compared to what the Company disclosed in its press release dated November 1, 2018.

Recently adopted accounting pronouncements

In June 2018, the Financial Accounting Standards Board ("FASB") issued Accounting Standards Update ("ASU") 2018-07, "Stock Compensation (Topic 718): Improvements to Nonemployee Share-Based Payment Accounting." ASU 2018-07 aligns the accounting for share-based payment awards issued to employees and nonemployees as well as improves financial reporting for share-based payments to nonemployees. The ASU is effective for fiscal years beginning after December 15, 2018, and interim periods within those fiscal years and will be applied to all new option awards granted after the date of adoption. Early adoption is permitted. The Company elected to early adopt ASU 2017-09 effective as of January 1, 2018 and applied it to share-based payment awards issued during the nine months ended September 30, 2018. The adoption of this standard did not have a material impact on the Company's consolidated financial statements and related disclosures.

In May 2017, the FASB issued ASU No. 2017-09, Compensation—Stock Compensation (Topic 718): Scope of Modification Accounting ("ASU 2017-09"), which clarifies when to account for a change to the terms or conditions of a share-based payment award as a modification. Under the new guidance, modification accounting is required only if the fair value, the vesting conditions or the classification of the award (as equity or liability) changes as a result of the change in terms or conditions. ASU 2017-09 is effective for annual periods beginning after December 15, 2017, including interim periods within those fiscal years. Early adoption is permitted. The Company adopted ASU 2017-09 as of the required effective date of January 1, 2018. The adoption of ASU 2017-09 will have an impact on the modification of stock-based awards, if any, after the date of adoption. The adoption of ASU 2017-09 did not have an impact on the Company's financial position, results of operations or cash flows.

In August 2016, the FASB issued ASU No. 2016-15, Statement of Cash Flows: Classification of Certain Cash Receipts and Cash Payments (Topic 230) ("ASU 2016-15"). This guidance addresses diversity in practice in how certain cash receipts and cash payments are presented in the statement of cash flows. The standard is

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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effective for public entities for fiscal years beginning after December 15, 2017, including interim periods in those fiscal years, and early adoption is permitted. The adoption of ASU 2016-15 is required to be applied retrospectively. The Company adopted ASU 2017-09 on the required effective date of January 1, 2018, and the adoption did not have an impact on the Company's financial position, results of operations or cash flows.

In May 2014, the FASB issued ASU No. 2014-09, Revenue from Contracts with Customers (Topic 606) ("ASU 2014-09"), which supersedes existing revenue recognition guidance under GAAP. The standard's core principle is that a company will recognize revenue when it transfers promised goods or services to customers in an amount that reflects the consideration to which the company expects to be entitled in exchange for those goods or services. The standard defines a five-step process to achieve this principle, and will require companies to use more judgment and make more estimates than under the current guidance. The Company expects that these judgments and estimates will include identifying performance obligations in the customer contract, estimating the amount of variable consideration to include in the transaction price and allocating the transaction price to each separate performance obligation. ASU 2014-09 also requires additional disclosure about the nature, amount, timing and uncertainty of revenue and cash flows arising from customer contracts. In August 2015, the FASB issued ASU No. 2015-14, Revenue from Contracts with Customers (Topic 606): Deferral of the Effective Date, which delays the effective date of ASU 2014-09 such that the standard is effective for public entities for annual periods beginning after December 15, 2017 and for interim periods within those fiscal years. Early adoption of the standard is permitted for annual periods beginning after December 15, 2016. The FASB subsequently issued amendments to ASU 2014-09 that have the same effective date and transition date. The Company adopted ASU 2014-09 as of the required effective date of January 1, 2018 and the adoption did not have an impact on the Company's consolidated financial statements as the Company does not currently have any revenue-generating arrangements.

Recently issued accounting pronouncements

In July 2017, the FASB issued ASU No. 2017-11*, Earnings Per Share (Topic 260), Distinguishing Liabilities from Equity (Topic 480), Derivatives and Hedging (Topic 815) (Part I) Accounting for Certain Financial Instruments with Down Round Features (Part II) Replacement of the Indefinite Deferral for Mandatorily Redeemable Financial Instruments of Certain Nonpublic Entities and Certain Mandatorily Redeemable Noncontrolling Interests with a Scope Exception ("ASU 2017-11").* Part I applies to entities that issue financial instruments such as warrants, convertible debt or convertible preferred stock that contain down-round features. Part II replaces the indefinite deferral for certain mandatorily redeemable noncontrolling interests and mandatorily redeemable financial instruments of nonpublic entities contained within ASC Topic 480 with a scope exception and does not impact the accounting for these mandatorily redeemable instruments. ASU 2017-11 is required to be adopted for annual periods beginning after December 15, 2018, including interim periods within those fiscal years. The Company is currently evaluating the impact that the adoption of ASU 2017-11 will have on its consolidated financial statements.

In February 2016, the FASB issued ASU No. 2016-02, *Leases (Topic 842)* ("ASU 2016-02"), which sets out the principles for the recognition, measurement, presentation and disclosure of leases for both parties to a

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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contract (i.e., lessees and lessors). The new standard requires lessees to apply a dual approach, classifying leases as either finance or operating leases based on the principle of whether or not the lease is effectively a financed purchase by the lessee. This classification will determine whether lease expense is recognized based on an effective interest method or on a straight-line basis over the term of the lease, respectively. A lessee is also required to record a right-of-use asset and a lease liability for all leases with a term of greater than 12 months regardless of their classification. Leases with a term of 12 months or less will be accounted for similar to existing guidance for operating leases today. The guidance is effective for public entities for annual periods beginning after December 15, 2018 and for interim periods within those fiscal years, and early adoption is permitted. The Company is still evaluating the full impact this standard will have on its consolidated financial statements and related disclosures, but expects to recognize substantially all of its leases on the balance sheet as of January 1, 2019, which is the Company's adoption date, by recording a right-of-use asset and a corresponding lease liability.

3. Fair value of financial assets and liabilities

Short-term investments as of September 30, 2018 consisted of U.S. Treasury notes all of which are due within six months. As of September 30, 2018, the fair value of short-term investments was $268,200 of which the amortized cost was $268,255 and gross unrealized loss was $55. The Company did not have any short-term investments as of December 31, 2017.

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

	Fair value measurements as of September 30, 2018 using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash—money market funds	$ 210	$ —	$—	$ 210
Cash equivalents—money market funds	5,299	—	—	5,299
Cash equivalents—U.S. Treasury notes	—	15,336	—	15,336
Short-term investments—U.S. Treasury notes	—	268,200	—	268,200
	$ 5,509	$ 283,536	$—	$ 289,045

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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	Fair value measurements as of December 31, 2017 using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Restricted cash—money market funds	$ 105	$ —	$—	$ 105
Cash equivalents—money market funds	5,487	—	—	5,487
Cash equivalents—U.S. Treasury notes	—	14,995	—	14,995
	$ 5,592	$ 14,995	$—	$ 20,587

During the periods ended September 30, 2018 and December 31, 2017 there were no transfers between Level 1, Level 2 and Level 3.

The money market funds were valued using quoted prices in active markets, which represent a Level 1 measurement in the fair value hierarchy. The Company's cash equivalents and short-term investments as of September 30, 2018 and cash equivalents as of December 31, 2017 also consisted of U.S. Treasury notes, which are not traded on a daily basis and, therefore, represent a Level 2 measurement in the fair value hierarchy at each period end.

4. Property and equipment, net

Property and equipment, net consisted of the following:

	September 30, 2018	December 31, 2017
Furniture and fixtures	$ 6	$ 83
Computer hardware and software	94	9
Vehicles	85	85
Construction in progress	2,241	—
	2,426	177
Less: Accumulated depreciation	(66)	(52)
	$ 2,360	$ 125

During the nine months ended September 30, 2018, the Company initiated the construction of a laboratory at Kiniksa US's headquarters. Construction in progress is primarily comprised of leasehold improvements and lab equipment which the Company anticipates will be placed into service by the end of 2018.

Depreciation expense was $12 and $8 during the three months ended September 30, 2018 and 2017, respectively, and $32 and $20 during the nine months ended September 30, 2018 and 2017, respectively.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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5. Accrued expenses

Accrued expenses consisted of the following:

	September 30, 2018	December 31, 2017
Accrued employee compensation and benefits	$ 3,160	$ 1,570
Accrued research and development expenses	9,313	3,905
Accrued legal and professional fees	548	688
Other	332	49
	$ 13,353	$ 6,212

6. 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 (deficit).

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 Company's 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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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and restated. In connection with the completion of its IPO in May 2018, the Company amended and restated its bye-laws (the "Amended & Restated Bye-Laws") to, among other things, authorize the issuance of undesignated preferred shares. As of September 30, 2018, 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 is 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 is 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.

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 is permitted by Bermuda law, into such number of fully paid and non-assessable Class B common shares as is determined by dividing the Series A Original Issue Price by the Series A Conversion Price (as defined below) in effect at the time of conversion. Each Series B preferred share 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 is permitted by Bermuda law, into such number of fully paid and non-assessable Class A common shares as is determined by dividing the Series B Original Issue Price by the Series B Conversion Price (as defined below) in effect at the time of conversion. Each Series C preferred share 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 is permitted by Bermuda law, into such number of fully paid and non-assessable Class A Shares as is determined by dividing the Series C Original Issue Price by the Series C Conversion Price (as defined below) in effect at the time of conversion.

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Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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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. The Series C Original Issue Price and Series C Conversion Price were equal to $15.6438. 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 is 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 is 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 elect 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 elect 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 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 receive, or simultaneously receive, 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 that is convertible into common shares, that dividend per Preferred Share as would equal the product of (a) the dividend payable on each share of such class or series determined, if applicable, as if all shares of such class or series had been converted into common shares and (b) the number of common shares issuable upon conversion of a share the applicable series of Preferred Shares, or (ii) in the case of a dividend on any class or series that is not convertible into common shares, at a rate per Preferred Share determined by (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. Prior to the Company's IPO, no cash dividends had been declared or paid.

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Notes to consolidated financial statements (Continued)

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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 shall be made to the holders of common shares by reason of their ownership thereof, an amount per share equal to the greater of (i) one times the applicable 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 be distributed among the holders of common shares, pro rata based on the number of shares held by each such holder.

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

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

Redemption

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

7. 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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Notes to consolidated financial statements (Continued)

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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 $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 of their over-allotment option to purchase additional shares at $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 Company's Amended & Restated Bye-Laws.

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. As of September 30, 2018, 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 or Class B1 common shares have no voting rights. As of December 31, 2017, the holders of the 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 the Class A and Class B common shares, voting together with the holders of the Preferred Shares, voting together as a single class, 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 board of directors. As of December 31, 2017, these dividends were subject to the preferential dividend rights of the holders of the Company's Preferred Shares. In May 2018, following the conversion of the Preferred Shares into common shares, these dividends are subject to the rights, powers and preferences of the preferred shares. As of September 30, 2018, no preferred shares were designated or issued. Through December 31, 2017 and September 30, 2018, 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 is convertible into one Class A common share or one Class B common share at the holder's election. There are no conversion rights associated with the Company's Class A common shares.

8. Share-based compensation

2018 Incentive award plan

In May 2018, the Company's board of directors and shareholders approved the 2018 Incentive Award Plan (the "2018 Plan"), which became effective on May 23, 2018. On the effectiveness of the 2018 Plan, the Company ceased granting awards under its 2015 Equity Incentive Plan (as amended, the "2015 Plan").

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. A total of 4,466,500 Class A common shares were initially reserved for issuance under the 2018 Plan. The number of Class A common shares that may be issued under the 2018 Plan will automatically increase on each January 1, beginning with the fiscal year ending December 31, 2019 and continuing for each fiscal year until, and including, the fiscal year ending December 31, 2028, equal to the lesser of (i) 4% of the Class A common shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (ii) a smaller number of Class A common shares determined by the Company's board of directors. No more than 27,915,000 Class A common shares may be issued under the 2018 Plan upon the exercise of incentive options. The Class A common shares underlying any awards issued under the 2018 Plan or the 2015 Plan that on or after the effective date of the 2018 Plan expire, lapse unexercised or are terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited under the 2018 Plan or the 2015 Plan will be added back to the Class A common shares available for issuance under the 2018 Plan. As of September 30, 2018, 3,280,059 shares remained available for future grant.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

2015 Equity incentive plan

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

As of December 31, 2017, the total number of Class A common shares authorized to be issued under the 2015 Plan was 4,794,266 shares and 1,644,893 shares were available for future grant. As of September 30, 2018, there were 4,620,850 shares of Class A common shares subject to outstanding awards under the plan authorized and reserved for issuance under the 2015 Plan pursuant such awards and no Class A common shares were otherwise authorized and reserved for issuance or available for future grant under the 2015 Plan as it was replaced by the 2018 Plan.

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

Shares that are expired, terminated, surrendered or canceled under the 2015 Plan without having been fully exercised will be available for future awards under the 2018 Plan.

Share option grants during the nine months ended September 30, 2018 and 2017

During the nine months ended September 30, 2018 and 2017, the Company granted options to purchase 2,887,639 and 1,434,156 Class A common shares, respectively, to employees and directors. The Company recorded share-based compensation expense for options granted to employees and directors of $1,414 and $321 during the three months ended September 30, 2018 and 2017, respectively, and $2,980 and $540 during the nine months ended September 30, 2018 and 2017, respectively.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

During the nine months ended September 30, 2018, the Company granted options to purchase 4,000 Class A common shares to non-employees. During the nine months ended September 30, 2017, the Company granted 1,829 options to purchase Class A common shares to non-employees. The Company recorded share-based compensation expense for options granted to non-employees of $35 and $1 during the three months ended September 30, 2018 and 2017, respectively, and $87 and $10 during the nine months ended September 30, 2018 and 2017, respectively.

2018 Employee share purchase plan

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

Option valuation

The assumptions that the Company used to determine the grant-date fair value of options granted to employees and directors under the 2015 Plan and the 2018 Plan (collectively, the "Plans") during the three and nine months ended September 30, 2018 and 2017 were as follows, presented on a weighted-average basis:

	Three months ended September 30,		Nine months ended September 30,	
	2018	2017	2018	2017
Risk-free interest rate	2.93%	1.90%	2.80%	1.97%
Expected term (in years)	6.25	6.00	6.41	6.00
Expected volatility	74.37%	73.88%	74.90%	74.23%
Expected dividend yield	0%	0%	0%	0%

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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

	Three months ended September 30,		Nine months ended September 30,	
	2018	2017	2018	2017
Risk-free interest rate	2.94%	1.97%	2.90%	1.97%
Expected term (in years)	7.04	8.84	7.46	8.84
Expected volatility	74.49%	77.93%	74.07%	77.93%
Expected dividend yield	0%	0%	0%	0%

Options

Share option activity under the Plans is summarized as follows:

	Number of shares	Weighted average exercise price	Weighted average remaining contractual term (in years)	Aggregate intrinsic value
Outstanding as of December 31, 2017	3,123,064	$ 2.75	8.82	$ 6,010
Granted	2,891,639	17.11		
Exercised	(22,060)	3.46		
Forfeited	(132,475)	5.16		
Outstanding as of September 30, 2018	5,860,168	$ 9.78	8.81	$ 96,055
Options exercisable as of September 30, 2018	1,575,324	$ 2.44	7.72	$ 36,330
Options unvested as of September 30, 2018	4,284,844	$ 12.48	9.22	$ 59,725

The aggregate intrinsic value of options is calculated as the difference between the exercise price of the options and the fair value of the Company's common shares for those options that had exercise prices lower than the fair value of the Company's common shares.

During the nine months ended September 30, 2018, option holders exercised 22,060 options for Class A common shares with an intrinsic value of $254 for total cash proceeds of $76.

The weighted-average grant-date fair value per share of options granted during the nine months ended September 30, 2018 and 2017 was $11.69 and $2.53, respectively.

The total fair value of options vested during the nine months ended September 30, 2018 and 2017 was $1,676 and $351, respectively.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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 nine months ended September 30, 2018:

	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, 2017	312,229	$ 0.000273235	1,635,495	$ 0.000273235
Granted	—	—	—	—
Vested	(133,812)	$ 0.000273235	(669,066)	$ 0.000273235
Unvested restricted shares outstanding as of September 30, 2018	178,417	$ 0.000273235	966,429	$ 0.000273235

The aggregate fair value of restricted shares that vested during the nine months ended September 30, 2018 and 2017 was $10,477 and $2,764, respectively.

Share-based compensation

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

	Three months ended September 30,		Nine months ended September 30,	
	2018	2017	2018	2017
Research and development expenses	$ 576	$ 121	$ 1,141	$ 181
General and administrative expenses	926	201	1,978	369
	$ 1,502	$ 322	$ 3,119	$ 550

As of September 30, 2018, total unrecognized compensation cost related to the unvested share-based awards was $34,676, which is expected to be recognized over a weighted average period of 3.54 years.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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9. License and acquisition agreements

Biogen asset purchase agreement

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to 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. During the year ended December 31, 2017, the Company made a milestone payment of $4,000 associated with the achievement of a specified clinical milestone event. Additionally, the Company could be obligated to make up to an aggregate of up to $150,000 of payments upon the achievement of specified annual net sales milestones and to pay tiered royalties 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 Company made insignificant payments in connection with the retained contracts during the three and nine months ended September 30, 2018 and 2017.

The Biogen Agreement will terminate upon the expiration of all payment obligations with respect to the last product in all countries in the territory. The Company has the right to terminate the agreement with 90 days' prior written notice. Both parties may terminate by mutual written consent or in the event of material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches).

During the three months ended September 30, 2018 and 2017 and the nine months ended September 30, 2018 and 2017, the Company recorded research and development expense of $11, $11, $41 and $4,158, respectively, in connection with milestone and other payments due under the Biogen Agreement.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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.

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 nine months ended September 30, 2018 and, as a result, this payment was made and is recorded in the Company's consolidated statement of operations for the nine months ended September 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 September 30, 2018 and December 31, 2017, the Company determined that the payment related to the milestone was not probable and, therefore, no amount was recorded in the Company's consolidated statement of operations and comprehensive loss during the nine months ended September 30, 2018 and 2017. The Company has also agreed to pay royalties on annual net sales of products licensed under the agreement.

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

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

During the three and nine months ended September 30, 2018, the Company recorded research and development expense of $4, and $154, respectively, in connection with milestone payments due under the Novo Nordisk Agreement. During the three and nine months ended September 30, 2017, the Company recorded research and development expense of $1,500, in connection with the upfront payment due under the Novo Nordisk Agreement.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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Primatope stock purchase option agreement

In September 2017, the Company entered into a stock purchase option agreement (the "Primatope Agreement") with Primatope Therapeutics, Inc. ("Primatope"), pursuant to which the Company has been granted a license to certain intellectual property rights controlled by Primatope to research, develop, and manufacture the pre-clinical antibody, KPL-404.

The agreement provides the Company with an exclusive call option to purchase 100% of the capital stock of Primatope. Upon execution of the agreement, the Company made $500 in upfront payments for the initial option period through April 2018 (the "Initial Option Period"). The Primatope Agreement allows up to three extensions of the Initial Option Period through January 2019 (including the initial option period, the "Option Period") for total extension payments of up to $800. Through October 2018, the Company made payments totaling $600 to extend the Option Period to November 15, 2018. During the Option Period, the Company may conduct research and pre-clinical work to assess the viability of the asset.

If the call option is exercised, the Company will acquire all of the outstanding equity of Primatope in exchange for upfront consideration of $10,000 as well as potential milestone payments of up to $10,000. The upfront payment and the milestone payments may be paid at the option of the Company in a combination of cash and issuance of the Company's Class A common shares.

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

Either party may terminate the Primatope Agreement for uncured material breach of the agreement by the other party or by mutual written consent.

During the three and nine months ended September 30, 2018, the Company recorded research and development expense of $250 and $500, respectively, related to the extension of the option period under the Primatope Agreement. During the three and nine months ended September 30, 2017, the Company recorded research and development expense of $500, in connection with upfront payments related to the Initial Option Period under the Primatope Agreement.

Regeneron license agreement

In September 2017, the Company entered into a license agreement (the "Regeneron Agreement") with Regeneron Pharmaceuticals, Inc. ("Regeneron"), pursuant to which the Company has been granted an exclusive, sublicensable license under certain intellectual property rights controlled by Regeneron to develop and commercialize 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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

business. The Company recorded the upfront payment as research and development expense in the consolidated statement of operations and comprehensive loss because the acquired technology represented in-process research and development and had no alternative future use.

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

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

The parties also entered into a clinical supply agreement under which Regeneron agreed to manufacture the developed product during the clinical phase. During the three and nine months ended September 30, 2018, the Company recorded research and development expense of $257 and $1,835, respectively, related to the purchase of drug materials under this agreement. During the three and nine months ended September 30, 2017, the Company did not incur any research and development expense related to the purchase of drug materials under this agreement. As of September 30, 2018 and December 31, 2017, the Company has non-cancelable purchase commitments under the clinical supply agreement (see Note 11).

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 if the products are determined to have certain safety concerns.

The Company did not incur any research and development expense directly related to milestone payments due under the Regeneron Agreement during the three and nine months ended September 30, 2018. During the three and nine months ended September 30, 2017, the Company recorded research and development expense of $5,000, in connection with the upfront payment 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

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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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 is anticipated to be 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 September 30, 2018 and December 31, 2017, the Company determined that the payment related to this milestone was probable and, therefore, recognized research and development expense and an accrued milestone of $10,000 during the year ended December 31, 2017. In addition, the Company is obligated to make clinical and regulatory milestone payments of up to $15,000 in the aggregate for each subsequent indication. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

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

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

During the three and nine months ended September 30, 2018 and 2017, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

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

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

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

	Three months ended September 30,		Nine months ended September 30,	
	2018	2017	2018	2017
Numerator:				
Net loss attributable to common shareholders	$ (24,406)	$ (16,029)	$ (60,647)	$ (32,172)
Denominator:				
Weighted average common shares outstanding—basic and diluted	48,183,424	1,942,106	23,174,841	1,675,133
Net loss per share attributable to common shareholders—basic and diluted	$ (0.51)	$ (8.25)	$ (2.62)	$ (19.21)

The Company's potentially dilutive securities, which include options, unvested restricted shares and convertible preferred shares, have been excluded from the computation of diluted net loss per share 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 because including them would have had an anti-dilutive effect:

	As of September 30,	
	2018	2017
Options to purchase common shares	5,860,168	3,012,176
Unvested restricted shares	1,144,846	2,215,351
Convertible preferred shares (as converted to common shares)	—	22,885,492
	7,005,014	28,113,019

Kiniksa Pharmaceuticals, Ltd.

Notes to consolidated financial statements (Continued)

(Amounts in thousands, except share and per share amounts)
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11. Commitments and contingencies

Lease agreements

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. The lease expires on July 31, 2021. Monthly lease payments include base rent as well as, ancillary charges such as the share of operating expenses and real estate taxes. Base rent is $73 for the remainder of 2018.

The following table summarizes the future minimum lease payments under non-cancelable operating lease commitments, for the Lexington office, as of September 30, 2018:

Year ending December 31,	
2018	$ 218
2019	872
2020	872
2021	508
	$ 2,470

The Company recognizes rent expense on a straight-line basis over the respective lease period and has recorded deferred rent for rent expense incurred but not yet paid. The Company recorded rent expense of $305 and $101 during the three months ended September 30, 2018 and 2017, respectively, and $714 and $301 during the nine months ended September 30, 2018 and 2017, respectively.

License agreements

The Company has entered into license agreements with various parties under which it is obligated to make contingent and non-contingent payments (see Note 9).

Manufacturing commitments

The Company entered into agreements with several contract manufacturing organizations to provide pre-clinical and clinical trial materials. As of September 30, 2018 and December 31, 2017, the Company had committed to minimum payments under these agreements totaling $34,891 and $7,766, respectively.

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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 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 September 30, 2018 or December 31, 2017.

Legal proceedings

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

2,654,984 Class A common shares