

**UNITED STATES  
SECURITIES AND EXCHANGE COMMISSION**

Washington, D.C. 20549

**FORM 10-Q**

(Mark One)

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

For the quarterly period ended June 30, 2018

OR

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

For the transition period from \_\_\_\_\_ to \_\_\_\_\_

Commission file number: 001-38492

**Kiniksa Pharmaceuticals, Ltd.**

(Exact name of registrant as specified in its charter)

**Bermuda**

(State or other jurisdiction of incorporation or organization)

**98-1327726**

(I.R.S. Employer Identification Number)

**Kiniksa Pharmaceuticals, Ltd.**

**Clarendon House**

**2 Church Street**

**Hamilton HM11, Bermuda**

**+1 (441) 295-5950**

(Address, zip code and telephone number, including area code of principal executive offices)

**Kiniksa Pharmaceuticals Corp.**

**100 Hayden Avenue**

**Lexington, MA, 02421**

**(781) 431-9100**

(Address, zip code and telephone number, including area code of agent for service)

**N/A**

(Former name, former address and former fiscal year, if changed since last report)

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

Indicate by check mark whether the registrant has submitted electronically and posted on its corporate Web site, if any, every Interactive Data File required to be submitted and posted pursuant to Rule 405 of Regulation S-T (§232.405 of this chapter) during the preceding 12 months (or for such shorter period that the registrant was required to submit and post such files). Yes  No

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

Large accelerated filer  Accelerated filer

Non-accelerated filer  (Do not check if a smaller reporting company) Smaller reporting company

Emerging growth company

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

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

As of July 31, 2018 there were 49,454,662 common shares outstanding in aggregate, comprised of:

15,762,235 Class A common shares, par value \$0.000273235 per share

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

12,995,954 Class A1 common shares, par value \$0.000273235 per share

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

**Kiniksa Pharmaceuticals, Ltd.****FORM 10-Q****FOR THE THREE AND SIX MONTHS ENDED JUNE 30, 2018****TABLE OF CONTENTS**

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**SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS**

This Quarterly Report on Form 10-Q, or this Quarterly Report, contains forward-looking statements. All statements other than statements of historical facts contained in this Quarterly Report, including statements regarding our future results of operations and financial position, business strategy, prospective products and product candidates, their expected properties, performance and impact on healthcare costs, the expected timeline for achievement of our clinical milestones, the timing of, and potential results from, clinical and other trials, marketing authorization from the U.S. Food and Drug Administration, or 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,” “target,” “project,” “contemplate,” “believe,” “estimate,” “predict,” “potential” or “continue” or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. The forward-looking statements in this Quarterly Report are only predictions. We have based these forward-looking statements largely on our current expectations and projections about future events and financial trends that we believe may affect our business, financial condition and results of operations. These forward-looking statements speak only as of the date of this Quarterly Report and are subject to a number of risks, uncertainties and assumptions, including those described under the sections in this Quarterly Report entitled “Risk Factors” and “Management’s Discussion and Analysis of Financial Condition and Results of Operations.” These forward-looking statements are subject to numerous risks, including, without limitation, the following:

- our status as a clinical-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;

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- 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
- ownership concentration in our executive officers and certain members of our senior management may prevent our shareholders from influencing significant corporate decisions.

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

You should read this Quarterly Report and any documents that we reference in this Quarterly Report that we have filed with the Securities and Exchange Commission, or SEC, 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.

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**Part I — Financial Information**

**Item 1. Financial Statements (unaudited)**

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

	<u>June 30, 2018</u>	<u>December 31, 2017</u>
<b>Assets</b>		
Current assets:		
Cash and cash equivalents	\$ 285,518	\$ 45,555
Restricted cash	263	105
Short-term investments	73,672	—
Prepaid expenses and other current assets	4,416	1,444
Total current assets	<u>363,869</u>	<u>47,104</u>
Property and equipment, net	350	125
Restricted cash	210	—
Deferred offering costs	—	25
Deferred tax assets	700	238
Total assets	<u>\$ 365,129</u>	<u>\$ 47,492</u>

**Liabilities, Convertible Preferred Shares and Shareholders' Equity (Deficit)**

Current liabilities:		
Accounts payable	\$ 561	\$ 1,218
Accrued expenses	12,583	6,212
Accrued milestone	10,000	10,000
Total current liabilities	23,144	17,430
Deferred rent	186	—
Total liabilities	23,330	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 June 30, 2018 and December 31, 2017, respectively; aggregate liquidation preference of \$0 and \$120,000 as of June 30, 2018 and December 31, 2017, respectively;	—	119,770
Shareholders' equity (deficit):		
Class A common shares, par value of \$0.000273235 per share; 15,754,771 shares and 719,976 shares issued and outstanding as of June 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 June 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 June 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 June 30, 2018 and December 31, 2017, respectively	4	—
Additional paid-in capital	469,018	1,289
Accumulated other comprehensive income	7	—
Accumulated deficit	(127,239)	(90,998)
Total shareholders' equity (deficit)	341,799	(89,708)
Total liabilities, convertible preferred shares and shareholders' equity (deficit)	\$ 365,129	\$ 47,492

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

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**KINIKSA PHARMACEUTICALS, LTD.**  
**CONSOLIDATED STATEMENTS OF OPERATIONS AND COMPREHENSIVE LOSS**  
(In thousands, except share and per share amounts)  
(Unaudited)

	Three Months Ended		Six Months Ended	
	June 30,		June 30,	
	2018	2017	2018	2017
Operating expenses:				
Research and development	\$ 17,200	\$ 9,272	\$ 29,831	\$ 12,417
General and administrative	4,327	2,120	8,036	4,022
Total operating expenses	21,527	11,392	37,867	16,439
Loss from operations	(21,527)	(11,392)	(37,867)	(16,439)
Interest income	1,066	153	1,371	226
Loss before provision for income taxes	(20,461)	(11,239)	(36,496)	(16,213)
Benefit (provision) for income taxes	202	36	255	70
Net loss and comprehensive loss	\$ (20,259)	\$ (11,203)	\$ (36,241)	\$ (16,143)
Net loss per share attributable to common shareholders—basic and diluted	\$ (0.97)	\$ (6.69)	\$ (3.09)	\$ (10.47)
Weighted average common shares outstanding—basic and diluted	20,787,288	1,673,500	11,735,578	1,541,691

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

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

	Convertible Preferred Shares		Common Shares		Additional Paid-In Capital	Accumulated OCI	Accumulated Deficit	Total Shareholders' (Deficit) Equity
	(Series A, B and C)		(Class A, B, A1 and B1)					
	Shares	Amount	Shares	Amount				
<b>Balances at December 31, 2017</b>	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	—	—	9,484,202	4	155,511	—	—	155,515

underwriting discounts and commissions and offering costs								
Exercise of options	—	—	4,574	—	17	—	—	17
Share-based compensation expense	—	—	—	—	1,617	—	—	1,617
Unrealized gain (loss) on short term investments	—	—	—	—	—	7	—	7
Net loss	—	—	—	—	—	—	(36,241)	(36,241)
<b>Balances at June 30, 2018</b>	<u>—</u>	<u>\$ —</u>	<u>49,447,198</u>	<u>\$ 13</u>	<u>\$469,018</u>	<u>\$ 7</u>	<u>\$ (127,239)</u>	<u>\$ 341,799</u>

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

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

	Six Months Ended June 30,	
	2018	2017
<b>Cash flows from operating activities:</b>		
Net loss	\$ (36,241)	\$ (16,143)
Adjustments to reconcile net loss to net cash used in operating activities:		
Depreciation expense	20	12
Share-based compensation expense	1,617	228
Loss on disposal of property and equipment	66	—
Non-cash rent expense	208	—
Deferred income taxes	(463)	(69)
Changes in operating assets and liabilities:		
Prepaid expenses and other current assets	(2,965)	(942)
Accounts payable	(747)	898
Accrued expenses	5,616	119
Net cash used in operating activities	<u>(32,889)</u>	<u>(15,897)</u>
<b>Cash flows from investing activities:</b>		
Purchases of property and equipment	(311)	(37)
Purchases of short-term investments	(73,676)	—
Net cash used in investing activities	<u>(73,987)</u>	<u>(37)</u>
<b>Cash flows from financing activities:</b>		
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	(2,825)	—
Proceeds from exercise of options	17	—
Net cash provided by financing activities	<u>347,207</u>	<u>39,873</u>
<b>Net increase (decrease) in cash and cash equivalents and restricted cash</b>	<b>240,331</b>	<b>23,939</b>
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	<u>\$ 285,991</u>	<u>\$ 80,014</u>
<b>Supplemental information:</b>		
Cash paid for income taxes	\$ 148	\$ —
<b>Supplemental disclosure of non-cash investing and financing activities:</b>		
Deferred offering costs included in accrued expenses and accounts payable	\$ 856	\$ —

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

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**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 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.

### ***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 June 30, 2018 and during the three and six months then ended, the Company was not the primary beneficiary of a VIE.

### ***Use of Estimates***

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

### ***Unaudited Interim Consolidated Financial Information***

The accompanying unaudited consolidated financial statements have been prepared in accordance with GAAP for interim financial information. The accompanying unaudited consolidated financial statements do not include all of the information and footnotes required by GAAP for complete consolidated financial statements. The accompanying year-end consolidated balance sheet was derived from audited financial statements, but does not include all disclosures required by GAAP. The unaudited interim consolidated financial statements have been prepared on the same basis as the audited annual consolidated financial statements and, in the opinion of management, reflect all adjustments, which include only normal recurring adjustments, necessary for the fair statement of the Company's financial position as of June 30, 2018 and the results of its operations for the three and six months ended June 30, 2018 and 2017 and its cash flows for the six months ended June 30, 2018 and 2017. The results for the three and six months ended June 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.

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

#### ***Reverse Stock Split***

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

#### ***Initial Public Offering***

On May 23, 2018, the Company's registration statement on Form S-1 relating to its initial public offering of its Class A common shares (the "IPO") was declared effective by the Securities and Exchange Commission ("SEC"). On May 29, 2018, the Company completed the IPO of 8,477,777 Class A common shares at \$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 \$155,515 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 there are conditions and events, considered in the aggregate, that raise substantial doubt about the Company's ability to continue as a going concern within one year after the date the consolidated financial statements are issued. As of June 30, 2018, the Company had an accumulated deficit of \$127,239. During the six months ended June 30, 2018, the Company incurred a net loss of \$36,241 and used \$32,889 of net cash in operating activities. The Company expects to continue to generate operating losses for the foreseeable future. As of June 30,

2018, the Company had cash, cash equivalents and short-term investments of \$359,190. 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.

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

**2. Summary of Significant Accounting Policies**

***Cash and Cash Equivalents***

The Company classifies deposits in banks, money market funds and cash invested temporarily in various instruments with maturities of three months or less at the time of purchase as cash and cash equivalents. At June 30, 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.

***Concentrations of Credit Risk***

Financial instruments that potentially subject the Company to concentrations of credit risk consist principally of cash, cash equivalents and short-term investments. At June 30, 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 June 30, 2018 and 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 June 30, 2018, the underlying cash balance of \$210 securing this letter of credit, was classified as non-current in the 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.

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

The Company's restricted cash, which is held in a money market fund, is carried at fair value, determined based on Level 1 inputs in the fair value hierarchy described above (see Note 3). The Company's cash equivalents and short-term investments, consisting of money market funds and U.S. Treasury notes, are carried at fair value, determined based on Level 1 and 2 inputs in the fair value hierarchy described above (see Note 3). The carrying values of the Company's 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 clinical development activities and clinical trials as well as to manufacture clinical trial materials. Non-refundable prepayments for goods or services that will be used or rendered for future research and development activities are recorded as prepaid expenses. Such amounts are recognized as an expense as the goods are delivered or the related services are performed, or until it is no longer expected that the goods will be delivered or the services rendered.

### ***Research Contract Costs and Accruals***

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

### ***Share-Based Compensation***

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

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

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

The fair value of each restricted share award is estimated on the date of grant based on the fair value of the Company's Class A 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,

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

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 May 23, 2018, when the Company's convertible preferred shares converted to common shares, the Company's convertible preferred shares contractually entitled the holders of such shares to participate in dividends but did not contractually require the holders of such shares to participate in losses of the Company. Accordingly, for periods in which the Company reported a net loss attributable to common shareholders, such losses were not allocated to convertible preferred shareholders. In periods in which the Company reports a net loss attributable to common shareholders, diluted net loss per share attributable to common shareholders is the same as basic net loss per share attributable to common shareholders, since dilutive common shares are not assumed to have been issued if their effect is anti-dilutive. The Company reported a net loss attributable to common shareholders for the three and six months ended June 30, 2018 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 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 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.

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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 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 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:

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	Fair Value Measurements as of June 30, 2018 Using:			
	Level 1	Level 2	Level 3	Total
<b>Assets:</b>				
Restricted cash - money market funds	\$ 473	\$ —	\$ —	\$ 473
Cash equivalents - money market funds	185,197	—	—	185,197
Cash equivalents - U.S. Treasury notes	—	58,949	—	58,949
Short-term investments - U.S. Treasury notes	—	73,672	—	73,672
	<u>\$ 185,670</u>	<u>\$ 132,621</u>	<u>\$ —</u>	<u>\$ 318,291</u>

	Fair Value Measurements as of December 31, 2017 Using:			
	Level 1	Level 2	Level 3	Total
<b>Assets:</b>				
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
	<u>\$ 5,592</u>	<u>\$ 14,995</u>	<u>\$ —</u>	<u>\$ 20,587</u>

During the periods ended June 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 June 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:

	June 30, 2018	December 31, 2017
Furniture and fixtures	\$ 6	\$ 83
Computer hardware and software	93	9
Vehicles	85	85
Construction in progress	219	—
	<u>403</u>	<u>177</u>
Less: Accumulated depreciation	(53)	(52)
	<u>\$ 350</u>	<u>\$ 125</u>

Depreciation expense was \$12 and \$6 during the three months ended June 30, 2018 and 2017, respectively, and \$20 and \$12 during the six months ended June 30, 2018 and 2017, respectively.

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#### 5. Accrued Expenses

Accrued expenses consisted of the following:

	June 30, 2018	December 31, 2017
Accrued employee compensation and benefits	\$ 2,158	\$ 1,570
Accrued research and development expenses	9,091	3,905
Accrued legal and professional fees	1,267	688
Other	67	49
	<u>\$ 12,583</u>	<u>\$ 6,212</u>

#### 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 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 June 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.

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#### *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.

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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#### *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.

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 \$155,515 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 June 30, 2018 no preferred shares were designated or issued.

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*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.

*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. As of June 30, 2018, no preferred shares were designated or issued. Through December 31, 2017 and June 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 June 30, 2018, 4,352,676 shares remained available for future grant.

***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.

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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 June 30, 2018, there were 4,682,066 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.

#### **Stock Option Grants During the Six Months Ended June 30, 2018 and 2017**

During the six months ended June 30, 2018 and 2017, the Company granted options to purchase 1,775,292 and 1,355,471 Class A common shares, respectively, to employees and directors. The Company recorded share-based compensation expense for options granted to employees and directors of \$1,021 and \$109 during the three months ended June 30, 2018 and 2017, respectively, and \$1,557 and \$219 during the six months ended June 30, 2018 and 2017, respectively.

During the six months ended June 30, 2018 and 2017, the Company did not grant options to purchase Class A common shares to non-employees. The Company recorded share-based compensation expense for options granted to non-employees of \$38 and \$6 during the three months ended June 30, 2018 and 2017, respectively, and \$61 and \$9 during the six months ended June 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 six months ended June 30, 2018 and 2017 were as follows, presented on a weighted-average basis:

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	Three Months Ended June 30,		Six Months Ended June 30,	
	2018	2017	2018	2017
Risk-free interest rate	2.87%	2.48%	2.72%	1.98%
Expected term (in years)	5.95	6.00	6.51	6.00
Expected volatility	74.83%	74.26%	75.22%	74.25%
Expected dividend yield	0%	0%	0%	0%

The assumptions that the Company used to determine the grant-date fair value of options granted to non-employees were as follows, presented on a weighted-average basis:

	Three Months Ended June 30,		Six Months Ended June 30,	
	2018	2017	2018	2017
Risk-free interest rate	2.98%	1.97%	2.88%	1.97%
Expected term (in years)	7.57	8.84	7.70	8.84
Expected volatility	73.69%	77.93%	73.83%	77.93%
Expected dividend yield	0%	0%	0%	0%

#### **Options**

Stock option activity under the Plans is summarized as follows:

Number of	Weighted	Weighted	Aggregate
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	Shares	Average Exercise Price	Average Remaining Contractual Term (in years)	Intrinsic Value
Outstanding as of December 31, 2017	3,123,064	\$ 2.75	8.82	\$ 6,010
Granted	1,775,292	10.89		
Exercised	(4,574)	3.80		
Forfeited	(88,745)	4.49		
Outstanding as of June 30, 2018	<u>4,805,037</u>	\$ 5.72	8.81	\$ 55,939
Options exercisable as of June 30, 2018	1,375,448	\$ 2.23	7.91	\$ 20,799
Options unvested as of June 30, 2018	3,429,589	\$ 7.13	9.16	\$ 35,140

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 six months ended June 30, 2018, an option holder exercised 4,574 options for Class A common shares with an intrinsic value of \$30 for total cash proceeds of \$17.

The weighted-average grant-date fair value per share of options granted during the six months ended June 30, 2018 and 2017 was \$7.51 and \$2.51, respectively.

The total fair value of options vested during the six months ended June 30, 2018 and 2017 was \$1,053 and \$173, respectively.

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**Restricted Shares**

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

The following table summarizes restricted share activity for the six months ended June 30, 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	(89,208)	\$ 0.000273235	(446,044)	\$ 0.000273235
Unvested restricted shares outstanding as of June 30, 2018	<u>223,021</u>	\$ 0.000273235	<u>1,189,451</u>	\$ 0.000273235

The aggregate fair value of restricted shares that vested during the six months ended June 30, 2018 and 2017 was \$5,239 and \$1,349, respectively.

**Share-Based Compensation**

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

	Three Months Ended June 30,		Six Months Ended June 30,	
	2018	2017	2018	2017
Research and development expenses	\$ 363	\$ 30	\$ 565	\$ 60
General and administrative expenses	696	84	1,052	168
	<u>\$ 1,059</u>	<u>\$ 114</u>	<u>\$ 1,617</u>	<u>\$ 228</u>

As of June 30, 2018, total unrecognized compensation cost related to the unvested share-based awards was \$16,609, which is expected to be recognized over a weighted average period of 4.3 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

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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 six months ended June 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 June 30, 2018 and 2017 and the six months ended June 30, 2018 and 2017, the Company recorded research and development expense of \$19, \$4,086, \$30 and \$4,147, respectively, in connection with milestone and other payments due under the Biogen Agreement.

***Novo Nordisk License Agreement***

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

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. The technology was transferred during the six months ended June 30, 2018 and, as a result, this payment was made and is recorded in the Company's statement of operations for the six months ended June 30, 2018. In addition, the Company is obligated to make milestone payments upon the achievement of specified clinical, regulatory and initial sales milestones and upon the achievement of annual net sales thresholds, including a payment of \$1,000 upon the earlier to occur of a specified regulatory milestone and January 2020, unless the Novo Nordisk Agreement is earlier terminated by either party. As of June 30, 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 six months ended June 30, 2018 and 2017. The Company has also agreed to pay royalties on annual net sales of products licensed under the agreement.

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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 six months ended June 30, 2018, the Company recorded research and development expense of \$150, in connection with milestone payments due under the Novo Nordisk Agreement. The Company did not incur any research and development expense directly related to milestone payments due under the Novo Nordisk Agreement during the three months ended June 30, 2018 or during the three and six months ended June 30, 2017.

#### ***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. In April and July 2018, the Company made payments totalling \$500 to extend the Option Period to October 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 six months ended June 30, 2018 the Company recorded research and development expense of \$250, related to the extension of the option period under the Primatope Agreement. The Company did not incur any research and development expense directly related to the Primatope Agreement during the three and six months ended June 30, 2017.

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

#### ***Regeneron License Agreement***

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

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

Under the Regeneron Agreement, the Company is also obligated to make payments to Regeneron of up to an aggregate of \$27,500 upon the achievement of specified regulatory milestones. Upon commercialization of the licensed products, the parties will share profits equally, 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 three and six months ended June 30, 2018, the Company recorded research and development expense of \$1,201 and \$1,577, respectively, related to the purchase of drug materials under this agreement. During the three and six months ended June 30, 2017, the Company did not incur any research and development expense related to the purchase of drug materials under this agreement. As of June 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 year’s written notice if the Company terminates the agreement following U.S. marketing approval of a rilonacept product developed by the Company. The Company may also terminate the agreement with three 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 six months ended June 30, 2017 and 2018.

### **MedImmune License Agreement**

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

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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 June 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, including a \$5,000 pass-through payment due upon the achievement of a specified clinical milestone event which may be met in the second half of 2018. The Company is obligated to make milestone payments to MedImmune of up to \$85,000 upon the achievement of annual net sales thresholds up to, but excluding, \$1,000,000 in annual net sales as well as additional milestone payments aggregating up to \$1,100,000 upon the achievement of additional specified annual net sales thresholds starting at \$1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

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

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

During the three and six months ended June 30, 2018 and 2017, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

## **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 June 30,		Six Months Ended June 30,	
	2018	2017	2018	2017
<b>Numerator:</b>				
Net loss attributable to common shareholders	\$ (20,259)	\$ (11,203)	\$ (36,241)	\$ (16,143)
<b>Denominator:</b>				
Weighted average common shares outstanding—basic and diluted	20,787,288	1,673,500	11,735,578	1,541,691
Net loss per share attributable to common shareholders— basic and diluted	\$ (0.97)	\$ (6.69)	\$ (3.09)	\$ (10.47)

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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 June 30,	
	2018	2017
Options to purchase common shares	4,805,037	2,933,491
Unvested restricted shares	1,412,472	2,482,977
Convertible preferred shares (as converted to common shares)	—	22,885,492
	6,217,509	28,301,960

## 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, total \$27. As of June 30, 2018, future minimum lease payments under non-cancelable operating lease commitments for the Wellesley Hills office, which are all due during the year ending December 31, 2018, totaled \$54.

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. Upon execution of the lease, the Company made a prepayment of \$67 for August 2018 base rent. Monthly lease payments begin in September 2018 and include base rent of approximately \$70 as well as, ancillary charges such as the share of operating expenses and real estate taxes.

The following table summarizes the future minimum lease payments under non-cancelable operating lease commitments, for the Lexington office, as of June 30, 2018:

Year Ending December 31,	
2018	\$ 285
2019	872
2020	872
2021	508
	\$ 2,537

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 \$276 and \$97 during

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the three months ended June 30, 2018 and 2017, respectively, and \$403 and \$194 during the six months ended June 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 June 30, 2018 and December 31, 2017, the Company had committed to minimum payments under these agreements totaling \$13,375 and \$7,766, respectively.

### *Indemnification Agreements*

In the ordinary course of business, the Company may provide indemnification of varying scope and terms to vendors, lessors, business partners and other parties with respect to certain matters including, but not limited to, losses arising out of breach of such agreements or from intellectual property infringement claims made by third parties. In addition, the Company has entered into indemnification agreements with members of its board of directors and officers that will require the Company, among other things, to indemnify them against certain liabilities that may arise by reason of their status or service as directors or officers. The maximum potential amount of future payments the Company could be required to make under these indemnification agreements is, in many cases, unlimited. To date, the Company has not incurred any material costs as a result of such indemnifications. The Company does not believe that the outcome of any claims under indemnification arrangements will have a material effect on its financial position, results of operations or cash flows, and it did not accrue any liabilities related to such obligations in its consolidated financial statements as of June 30, 2018 or December 31, 2017.

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

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

*The following discussion and analysis of our financial condition and results of operations should be read in conjunction with our unaudited consolidated financial statements and related notes appearing elsewhere in this Quarterly Report and our audited consolidated financial statements and related notes for the year ended December 31, 2017 included in our final prospectus for the initial public offering of our common shares, or the IPO, filed with the Securities and Exchange Commission, or the SEC, pursuant to Rule 424(b)(4) on May 24, 2018. Some of the information contained in this discussion and analysis or set forth elsewhere in this Quarterly Report, including information with respect to our plans and strategy for our business, includes forward-looking statements within the meaning of Section 27A of the Securities Act of 1933, as amended, and Section 21E of the Securities Exchange Act of 1934, as amended, that involve risks and uncertainties. As a result of many factors, including those factors set forth in the risks identified in Part II-Item 1A “Risk Factors” section of this Quarterly Report and our other filings with the SEC, our actual results could differ materially from the results, performance or achievements expressed in or implied by these forward-looking statements.*

**Overview**

We are a clinical-stage biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. We have a pipeline of product candidates across various stages of development, currently focused on autoinflammatory and autoimmune conditions. We have three clinical-stage product candidates, one of which is anticipated to commence a Phase 3 clinical trial in 2018. We follow a disciplined and methodical approach to selectively identify 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.

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. 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 had 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 the 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 \$36.2 million and \$16.1 million for the six months ended June 30, 2018 and 2017, respectively. As of June 30, 2018, we had an accumulated deficit of \$127.2 million. We expect to continue to incur significant operating losses for at least the next several years as we advance our product candidates through all stages of development and clinical trials and, ultimately, seek regulatory approval. In addition, if we obtain marketing approval for any of our product candidates, we expect to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates. We expect to continue to incur additional costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses that we did not incur as a private company.

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

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

As of June 30, 2018, we had cash, cash equivalents and short-term investments of \$359.2 million. We believe that our existing cash, cash equivalents and short-term investments as of June 30, 2018 will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months from the date of issuance of the unaudited consolidated financial statements included in this Quarterly Report. We have based this estimate on assumptions

that may prove to be wrong, and we could exhaust our available capital resources sooner than we expect. See “— Liquidity and Capital Resources.” Our future viability beyond that point is dependent on our ability to raise additional capital to finance our operations.

## Components of our Results of Operations

### Revenue

To date, we have not generated any revenue from product sales and do not expect to generate any revenue from the sale of products in the foreseeable future. If our development efforts for our product candidates are successful and result in regulatory approval, we may generate revenue in the future from product sales.

### Operating Expenses

#### Research and Development Expenses

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

- expenses incurred to conduct the necessary 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;

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- 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:

	Three Months Ended June 30,		Six Months Ended June 30,	
	2018	2017	2018	2017
	(in thousands)		(in thousands)	
Riloncept	\$ 2,903	\$ —	\$ 4,126	\$ —
Mavrilimumab	2,465	—	2,743	—
KPL-716 (1)	6,712	8,045	13,928	10,246
KPL-045 (2)	890	—	1,327	—
KPL-404 (3)	671	—	1,046	—
Unallocated research and development expenses	3,559	1,227	6,661	2,171
<b>Total research and development expenses</b>	<b>\$ 17,200</b>	<b>\$ 9,272</b>	<b>\$ 29,831</b>	<b>\$ 12,417</b>

(1) The amount for the three and six months ended June 30, 2017 includes expense of \$4.1 million related to a milestone payment under our asset purchase agreement with Biogen associated with the achievement of a specified clinical milestone event.

(2) The amount for the six months ended June 30, 2018 includes expense of \$0.2 million related to a technology transfer payment under our license agreement with Novo Nordisk.

(3) The amount for the three and six months ended June 30, 2018 includes expense of \$0.3 million related to the extension of the option period 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

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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 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

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

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 Three Months Ended June 30, 2018 and 2017

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

	Three Months Ended June 30,		Change
	2018	2017	
(in thousands)			
<b>Operating expenses:</b>			
Research and development	\$ 17,200	\$ 9,272	\$ 7,928
General and administrative	4,327	2,120	2,207
Total operating expenses	<u>21,527</u>	<u>11,392</u>	<u>10,135</u>
Loss from operations	(21,527)	(11,392)	(10,135)
Interest income	1,066	153	913
Loss before provision for income taxes	(20,461)	(11,239)	(9,222)
Benefit (provision) for income taxes	202	36	166
Net loss	<u>\$ (20,259)</u>	<u>\$ (11,203)</u>	<u>\$ (9,056)</u>

### Research and Development Expenses

	Three Months Ended June 30,		Change
	2018	2017	
(in thousands)			
<b>Direct research and development expenses by program:</b>			
Rilonacept	\$ 2,903	\$ —	\$ 2,903
Mavrilimumab	2,465	—	2,465
KPL-716	6,712	8,045	(1,333)
KPL-045	890	—	890
KPL-404	671	—	671
<b>Unallocated research and development expenses:</b>			
Personnel related (including share-based compensation)	2,816	945	1,871
Other	743	282	461
Total research and development expenses	<u>\$ 17,200</u>	<u>\$ 9,272</u>	<u>\$ 7,928</u>

Research and development expenses were \$17.2 million for the three months ended June 30, 2018, compared to \$9.3 million for the three months ended June 30, 2017. The increase of \$7.9 million was primarily due to an increase in external fees related to our development programs, of which there were five in 2018 and one in 2017, as well as an increase of \$2.3 million in unallocated research and development expenses.

The direct costs of \$2.9 million for our rilonacept program during the three months ended June 30, 2018 were due to expenses related to clinical research and development with our open-label Phase 2 proof-of-concept clinical

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trial and preparation for our planned Phase 3 clinical trial. We had no direct costs for our rilonacept program during the three months ended June 30, 2017.

The direct costs of \$2.5 million for our mavrilimumab program during the three months ended June 30, 2018 were due to expenses related primarily to preparation for our planned clinical trials, including a Phase 2 trial in giant cell arteritis. We had no direct costs for our mavrilimumab program during the three months ended June 30, 2017.

The direct costs of our KPL-716 program were \$6.7 million during the three months ended June 30, 2018, compared to \$8.0 million during the three months ended June 30, 2017. The decrease in \$1.3 million in direct costs for our KPL-716 program during the three months ended June 30, 2018 was primarily due to expense of \$4.1 million in the three months ended June 30, 2017 related to a milestone payment associated with the achievement of a specified clinical milestone event under our asset purchase agreement with Biogen MA Inc., or Biogen, partially offset by an increase in manufacturing and development costs related to clinical drug supply and our Phase 1a/1b clinical trial.

The direct costs of \$0.9 million for our KPL-045 program during the three months ended June 30, 2018 were due to direct costs related to clinical research and development, including manufacturing development costs. We had no direct costs for our KPL-045 program during the three months ended June 30, 2017.

The direct costs of \$0.7 million for our KPL-404 program during the three months ended June 30, 2018 were due to \$0.4 million of direct costs related to clinical research and development, including manufacturing development costs as well as \$0.3 million related to the extension of the option period under our stock purchase option agreement with Primatope Therapeutics, Inc., or Primatope. We had no direct costs for our KPL-404 program during the three months ended June 30, 2017.

Unallocated research and development expenses were \$3.6 million for the three months ended June 30, 2018 compared to \$1.2 million for the three months ended June 30, 2017. The increase of \$2.4 million in unallocated research and development expenses was due to an increase of \$1.9 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 process development and manufacturing of drug supply and coordinating with CROs on the conduct and oversight of our current and planned clinical trials as well as research studies and development programs for our product candidates. Personnel-related costs for the three months ended June 30, 2018 and 2017 included share-based compensation of \$0.4 million and \$30,000, respectively.

## General and Administrative Expenses

General and administrative expenses were \$4.3 million for the three months ended June 30, 2018 compared to \$2.1 million for the three months ended June 30, 2017. The increase of \$2.2 million was due to increases of \$1.5 million in personnel-related costs and \$0.7 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, finance and human resources departments, as we continued to expand our operations to support the organization. Personnel-related costs for the three months ended June 30, 2018 and 2017 included share-based compensation of \$0.7 million and \$0.1 million, respectively. Professional fees increased due to legal costs incurred in connection with maintaining and registering worldwide patents and costs associated with our ongoing business operations, as well as higher accounting, recruiting, market research expenses and other costs incurred due to becoming a public company.

## Interest Income

Interest income was \$1.1 million for the three months ended June 30, 2018 compared to \$0.2 million for the three months ended June 30, 2017. The increase was due to both higher average invested balances and higher interest rates on U.S. Treasury notes in 2018.

## Benefit (Provision) for Income Taxes

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

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### Comparison of the Six Months Ended June 30, 2018 and 2017

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

	Six Months Ended June 30,		Change
	2018	2017	
	(in thousands)		
Operating expenses:			
Research and development	\$ 29,831	\$ 12,417	\$ 17,414
General and administrative	8,036	4,022	4,014
Total operating expenses	<u>37,867</u>	<u>16,439</u>	<u>21,428</u>
Loss from operations	(37,867)	(16,439)	(21,428)
Interest income	1,371	226	1,145
Loss before provision for income taxes	(36,496)	(16,213)	(20,283)
Benefit (provision) for income taxes	255	70	185
Net loss	<u>\$ (36,241)</u>	<u>\$ (16,143)</u>	<u>\$ (20,098)</u>

## Research and Development Expenses

	Six Months Ended June 30,		Change
	2018	2017	
	(in thousands)		
Direct research and development expenses by program:			
Rilonacept	\$ 4,126	\$ —	\$ 4,126
Mavrilimumab	2,743	—	2,743
KPL-716	13,928	10,246	3,682
KPL-045	1,327	—	1,327
KPL-404	1,046	—	1,046
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	5,273	1,683	3,590
Other	1,388	488	900
Total research and development expenses	<u>\$ 29,831</u>	<u>\$ 12,417</u>	<u>\$ 17,414</u>

Research and development expenses were \$29.8 million for the six months ended June 30, 2018, compared to \$12.4 million for the six months ended June 30, 2017. The increase of \$17.4 million was primarily due to an increase in external fees related to our development programs, of which there were five in 2018 and one in 2017, as well as an increase of \$4.5 million in unallocated research and development expenses.

The direct costs of \$4.1 million for our rilonacept program during the six months ended June 30, 2018 were due to expenses related to clinical research and development with our open-label Phase 2 proof-of-concept clinical trial and preparation for our planned Phase 3 clinical trial. We had no direct costs for our rilonacept program during the six months ended June 30, 2017.

The direct costs of \$2.7 million for our mavrilimumab program during the six months ended June 30, 2018 were due to expenses related primarily to preparation for our planned clinical trials including a Phase 2 trial in giant cell arteritis. We had no direct costs for our mavrilimumab program during the six months ended June 30, 2017.

The direct costs of our KPL-716 program were \$13.9 million during the six months ended June 30, 2018, compared to \$10.2 million during the six months ended June 30, 2017. The increase of \$3.7 million in direct costs for our KPL-716 program during the six months ended June 30, 2018 was primarily due to increased expenses

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related to manufacturing and development costs related to clinical drug supply and our Phase 1a/1b clinical trial.

The direct costs of \$1.3 million for our KPL-045 program during the six months ended June 30, 2018 were due to \$1.1 million of direct costs related to clinical research and development, including manufacturing development costs as well as \$0.2 million related to a technology transfer payment under our license agreement with Novo Nordisk A/S, or NovoNordisk. We had no direct costs for our KPL-045 program during the six months ended June 30, 2017.

The direct costs of \$1.0 million for our KPL-404 program during the six months ended June 30, 2018 were due to \$0.7 million of direct costs related to clinical research and development, including manufacturing development costs as well as \$0.3 million related to the extension of the option period under our stock purchase option agreement with Primatope. We had no direct costs for our KPL-404 program during the six months ended June 30, 2017.

Unallocated research and development expenses were \$6.7 million for the six months ended June 30, 2018 compared to \$2.2 million for the six months ended June 30, 2017. The increase of \$4.5 million in unallocated research and development expenses was due to an increase of \$3.6 million in personnel-related costs, including share-based compensation, and an increase of \$0.9 million in other costs. The increase in personnel-related costs was primarily due to the hiring of additional personnel in our research and development functions, particularly those responsible for coordinating with CMOs on process development and manufacturing of drug supply and coordinating with CROs on the conduct and oversight of our current and planned clinical trials as well as research studies and development programs for our product candidates. Personnel-related costs for the six months ended June 30, 2018 and 2017 included share-based compensation of \$0.6 million and \$0.1 million, respectively.

#### *General and Administrative Expenses*

General and administrative expenses were \$8.0 million for the six months ended June 30, 2018 compared to \$4.0 million for the six months ended June 30, 2017. The increase of \$4.0 million was due to increases of \$2.3 million in personnel-related costs and \$1.7 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, finance and human resources departments, as we continued to expand our operations to support the organization. Personnel-related costs for the six months ended June 30, 2018 and 2017 included share-based compensation of \$1.1 million and \$0.2 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 \$1.4 million for the six months ended June 30, 2018 compared to \$0.2 million for the six months ended June 30, 2017. The increase was due to both higher average invested balances and higher interest rates on U.S. Treasury notes in 2018.

#### *Benefit (Provision) for Income Taxes*

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

### **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 had 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 the 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

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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 June 30, 2018, we had cash, cash equivalents and short-term investments of \$359.2 million.

#### **Cash Flows**

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

	Six Months Ended June 30,	
	2018	2017
	(in thousands)	
Net cash used in operating activities	\$ (32,889)	\$ (15,897)
Net cash used in investing activities	(73,987)	(37)
Net cash provided by financing activities	347,207	39,873
Net increase (decrease) in cash and cash equivalents and restricted cash	<u>\$ 240,331</u>	<u>\$ 23,939</u>

#### *Operating Activities*

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

During the six months ended June 30, 2017, operating activities used \$15.9 million of cash, primarily resulting from our net loss of \$16.1 million and \$0.1 million in net cash used in changes in our operating assets and liabilities, partially offset by non-cash charges of \$0.2 million.

#### *Investing Activities*

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

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

#### *Financing Activities*

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

During the six months ended June 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.

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#### ***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 of 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
- acquire or in-license other product candidates and technologies.

We believe that our existing cash, cash equivalents and short-term investments at June 30, 2018 will enable us to fund our operating expenses and capital expenditure requirements for at least the next twelve months from the issuance date of the unaudited consolidated financial statements appearing elsewhere in this Quarterly Report. 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;

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- 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 disclosure of our contractual obligations and commitments is set forth under the heading "Management's Discussion and Analysis of Financial Condition and Results of Operations—Contractual Obligations and Commitments" in our final prospectus filed with the SEC on May 24, 2018. See Note 11 to our consolidated financial statements included in Item 1, "Consolidated Unaudited Financial Statements," of this Quarterly Report on Form 10-Q for a discussion of obligations and commitments.

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

During the six months ended June 30, 2018, there were no material changes to our critical accounting policies. Our critical accounting policies are described under the heading "Management's Discussion and Analysis of Financial Condition and Results of Operations— Critical Accounting Policies and Significant Judgments and Estimates" in our final prospectus related to our Registration Statement filed on S-1 (File No: 333-224488) filed with the SEC on May 24, 2018 and the notes to the consolidated financial statements included in Item 1, "Consolidated Unaudited Financial Statements," included in this Quarterly Report on Form 10-Q. We believe that of our critical accounting policies, the following accounting policies involve the most judgment and complexity:

- accrued research and development expenses;
- share-based compensation; and
- determination of the fair value of common shares prior to the IPO.

### **Emerging Growth Company Status**

The Jumpstart Our Business Act, or 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 SEC.

### **Recently Issued Accounting Pronouncements**

A description of recently issued accounting pronouncements that may potentially impact our financial position and results of operations is provided in Note 2 to our consolidated financial statements appearing at the beginning of this Quarterly Report on Form 10-Q.

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## **Item 3. Quantitative and Qualitative Disclosures About Market Risk.**

### ***Interest Rate Risk***

We are exposed to market risk related to changes in interest rates. As of June 30, 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.

## **Item 4. Controls and Procedures.**

### ***Limitations on Effectiveness of Controls and Procedures***

In designing and evaluating our disclosure controls and procedures, management recognizes that any controls and procedures, no matter how well designed and operated, can provide only reasonable assurance of achieving the desired control objectives. In addition, the design of disclosure controls and procedures must reflect the fact that there are resource constraints and that management is required to apply judgment in evaluating the benefits of possible controls and procedures relative to their costs.

### ***Evaluation of Disclosure Controls and Procedures***

Our management, with the participation of our Chief Executive Officer and Chief Financial Officer, evaluated, as of the end of the period covered by this Quarterly Report, the effectiveness of our disclosure controls and procedures (as defined in Rules 13a-15(e) and 15d-15(e) under the Securities Exchange Act of 1934, as amended, or the Exchange Act). Based on that evaluation, our Chief Executive Officer and Chief Financial Officer concluded that our disclosure controls and procedures were effective at the reasonable assurance level as of June 30, 2018.

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

There was no change in our internal control over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act) that has materially affected, or is reasonably likely to materially affect, our internal control over financial reporting.

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## **PART II - OTHER INFORMATION**

### **Item 1. Legal Proceedings.**

We are not party to any material legal proceedings.

### **Item 1A. Risk Factors.**

*Investing in our Class A common shares involves a high degree of risk. You should consider carefully the risks described below, together with the other information included or incorporated by reference in this Quarterly Report. If any of the following risks occur, our business, financial condition, results of operations and future growth prospects could be materially and adversely affected. In these circumstances, the market price of our Class A common shares could decline.*

### **Risks Related to Our Financial Position and Capital Needs**

***We are a clinical-stage 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 initiate or successfully 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 six months ended June 30, 2018 and 2017 were \$36.2 million and \$16.1 million, respectively. As of June 30, 2018, we had an accumulated deficit of \$127.2 million. We expect to continue to incur losses for the foreseeable future, and we anticipate these losses will increase substantially as we:

- continue our research and pre-clinical and clinical development of our product candidates, including our ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept for the treatment of recurrent pericarditis, and our repeat single-dose 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 to advance to a Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, our plans to advance to a Phase 2 clinical trial with mavrilimumab for the treatment of giant cell arteritis, or GCA, and pending clinical data from the single-dose portion of our Phase 1a/1b clinical trial and our LOTUS-PN study, our plans to initiate a Phase 2 clinical trial with KPL-716 in prurigo nodularis;
- initiate additional pre-clinical studies and clinical trials for our product candidates;
- increase our manufacturing needs or add additional manufacturers or suppliers;

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- 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, acquire or develop additional product candidates;
- make milestone or other payments under any license or purchase agreements;
- seek to maintain, protect and expand our intellectual property portfolio;
- seek to attract and retain skilled personnel;
- create additional infrastructure to support our operations as a public company and our product development and planned future commercialization efforts; and
- 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 and, continue our ongoing and anticipate beginning new 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:

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- the results, time and cost necessary for completing our ongoing open-label Phase 2 proof-of-concept clinical trial for rilonacept for the treatment of recurrent pericarditis and our repeat single-dose portion of our ongoing Phase 1b clinical trial of KPL-716 in subjects with atopic dermatitis, as well as our plans for advancing the clinical development of our programs, including our plans to advance to a Phase 3 clinical trial with rilonacept for the treatment of recurrent pericarditis, our plans to advance to a Phase 2 clinical trial with mavrilimumab for the treatment of GCA, and pending clinical data from the single-dose portion of our Phase 1a/1b clinical trial and our LOTUS-PN study, our plans to initiate a Phase 2 clinical trial with KPL-716 in prurigo nodularis;
- 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 Pharmaceuticals, Inc., or Regeneron, MedImmune, Limited, or MedImmune, Biogen, 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 in connection with the exercise of our option to purchase 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 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.

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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, restrict our operations or require us to relinquish rights to our technologies or product candidates.***

Until such time as we can generate substantial product revenues, if ever, we expect to finance our cash needs through securities offerings or debt financings, or possibly, license and collaboration agreements or research grants. The terms of any financing may adversely affect the holdings or the rights of our shareholders and our issuance of additional securities, whether equity or debt, or the possibility of such issuance, may cause the market price of our Class A common shares to decline. The sale of additional equity or convertible securities would dilute all of our shareholders. The incurrence of indebtedness would result in increased fixed payment obligations and we may be required to agree to certain restrictive covenants, such as limitations on our ability to incur additional debt, limitations on our ability to acquire, sell or license intellectual property rights and other operating restrictions that could adversely impact our ability to conduct our business. We could also be required to seek funds through arrangements with collaborators or 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. None of them have been previously studied in the indications for which we are developing them. We may not be able to demonstrate that they are safe or effective in the indications for which we are studying them and they may not be approved. Although rilonacept is approved and marketed for human use for the treatment of cryopyrin-associated periodic syndromes, or CAPS, in the United States by Regeneron, we are studying rilonacept for the treatment of recurrent pericarditis in an open-label Phase 2 proof-of-concept clinical trial, and we plan to advance development to a Phase 3 clinical trial in 2018. Mavrilimumab has been through Phase 2 clinical trials conducted by MedImmune for the treatment of rheumatoid arthritis, or RA, but we plan to commence Phase 2 clinical trials of mavrilimumab for the treatment of GCA. Our third product candidate, KPL-716, is currently in the repeat single dose portion of our ongoing Phase 1b clinical trial in subjects with atopic dermatitis. 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. We also have pre-clinical product

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candidates that will need to progress through studies to enable an IND prior to clinical development. None of our product candidates have advanced into a pivotal study for the indications in which we are studying them. 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.

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 conduct of clinical trials by 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 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 continuing our ongoing open-label Phase 2 proof-of-concept clinical trial for riloncept for the treatment of recurrent pericarditis, and our repeat single-dose portion of our ongoing Phase 1b clinical trial of KPL-716 in subjects with atopic dermatitis. We also plan to advance to a Phase 3 clinical trial with riloncept for the treatment of recurrent pericarditis, advance to a Phase 2 clinical trial with mavrilimumab for the treatment of GCA, and pending clinical data from the single-dose portion of our Phase 1a/1b clinical trial and our LOTUS-PN study, plan to initiate a Phase 2 clinical trial with KPL-716 in prurigo nodularis. We are also continuing preparations for IND-enabling studies of 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, the FDA, European regulatory authorities or other 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. We believe

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that the FDA's communications with MedImmune suggest 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, which we are pursuing. However, the FDA may disagree and may require that we generate additional data, or that we implement additional monitoring or other trial design changes prior to initiating clinical trials of mavrilimumab in the United States.

Further, we could discover that our clinical trial design leads to enrollment difficulties which could require protocol amendments and further delay our study. 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 Institutional Review Board, or IRB, approval at each clinical trial site;
- delays in or failure to obtain regulatory approval to commence a trial, or imposition of a clinical hold by regulatory agencies, after review of an IND or IND amendment, or equivalent application or amendment, or an inspection of our clinical trial operations or study sites;
- challenges in recruiting and enrolling suitable patients to participate in our clinical trials;
- amendments to 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;

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- 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 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 our 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. 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 may 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 identify a third party to manufacture mavrilimumab for any Phase 3 clinical trials and commercialization efforts, if any, and will need to transfer the manufacturing process of mavrilimumab to such third-party CMO. 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 the mavrilimumab.

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 theoretical risk of serious infections and cancer. Some common side effects of rilonacept include, cold symptoms, nausea, stomach pain, diarrhea, numbness or tingling feeling and injection-site reaction. For mavrilimumab, there is a theoretical risk for the development of pulmonary alveolar proteinosis, or PAP. PAP is a rare lung disorder in which surfactant-derived lipoproteins accumulate excessively within pulmonary alveoli due to loss of GM-CSF function. The disease can range in severity from a sub-clinical reduction in diffusion capacity to significant dyspnea during mild exertion. In 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 trials reveal a high or unacceptable severity and prevalence of these or other side effects, the FDA or applicable foreign regulatory agency may not authorize us to initiate our trials, or if initiated, our clinical 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:

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- 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 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 licensed or acquired or may in the future 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. None of our product candidates have been studied 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 has not been determined in recurrent pericarditis or GCA, respectively, and each may fail to receive regulatory approval for those indications.

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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 bodies 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. 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

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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;
- 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 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

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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.

Riloncept 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 riloncept, 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 riloncept 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 intend to seek orphan drug designation for certain of our product candidates, such as rilonacept, 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 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 seek Orphan Drug Designation for our other 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 a 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 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.

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### **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 plan to build 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.

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. In addition, 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. 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. We also contract with Regeneron to produce riloncept, and with CMOs for the manufacture of KPL-716 drug substance and drug product. We contract with CMOs in connection with our planned production of our pre-clinical product candidates, KPL-045 and KPL-404; however, we plan to build small scale manufacturing capabilities to support certain pre-clinical and early clinical development for these product candidates.

The facilities used by our contract manufacturers 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 contract manufacturers for compliance with cGMPs and other regulatory requirements in connection with the manufacture of our product candidates. If our contract manufacturers 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 contract manufacturers 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.

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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 contract manufacturers 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 are building our own manufacturing facilities to support the early development of our product candidates, KPL-045 and KPL-404, and we may be unsuccessful in doing so in a timely manner, which could delay or prevent the commencement of our planned clinical studies for these product candidates.***

We are building small scale manufacturing capacity 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.

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***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 products 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 active pharmaceutical ingredient, drug product and drug substance 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 active pharmaceutical ingredients, or API, drug product and drug substance used in riloncept, mavrilimumab and KPL-716 are supplied to us from single-source suppliers. For example, although Regeneron has been producing riloncept 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 API, drug product and drug substance 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 API, drug product or drug substance in the event any of our current suppliers of such API, drug product and drug substance cease their operations or stop offering us sufficient quantities of these materials for any reason.

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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 riloncept, 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 riloncept, 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 API, drug product and drug substance 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 API, drug product and drug substance used in our product candidates, any interruption or delay in the supply of components or materials, or our inability to obtain such API, drug product and drug substance 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.

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***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, ClinicalTrials.gov, within certain timeframes. Failure to do so when required can result in fines, adverse publicity and civil and criminal sanctions.

Although we intend 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

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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 the 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 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 choose 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;

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- 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-1 $\alpha$  and IL-1 $\beta$ , anakinra (KINERET), and one currently marketed product that modulates the signaling of IL-1 $\beta$ , canakinumab

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(ILARIS). There are other therapies which modulate IL-1 $\alpha$  and IL-1 $\beta$  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 enter clinical trials in GCA in 2018. In addition, Eli Lilly and Company is conducting clinical trials in GCA for baricitinib, and Sanofi S.A. and Regeneron intend to initiate a Phase 3 clinical trial in GCA for sarilumab (KEVZARA) in 2018. 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 will also be 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

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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 will need 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 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 compete effectively will depend, in part, on our ability to effectively manage the future development and expansion of our company.

***We may not be successful in executing our growth strategy to identify, discover, develop, in-license or acquire additional product candidates or our growth strategy may not deliver the anticipated results.***

We plan to source new product candidates that are complementary to our existing product candidates through our internal discovery program, or in-licensing, entering into strategic transactions or otherwise acquiring them from other companies or academic institutions. If we are unable to identify, discover, develop, in-license or acquire and integrate product candidates in accordance with this strategy, our ability to pursue this part of our growth strategy would be limited.

Research programs and business development efforts to identify new product candidates require substantial technical, financial and human resources. We may focus our efforts and resources on potential programs or product candidates that ultimately prove to be unsuccessful. In-licensing and acquisitions of technology often require significant payments and expenses and will consume additional resources. We will need to devote a substantial amount of time and personnel to research, develop and commercialize any acquired technology, in addition to our existing portfolio of programs. Our research programs, business

development efforts or licensing attempts may fail to yield additional complementary or successful product candidates for clinical development and commercialization 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 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;
- for product candidates we seek to in-license or acquire, we may not be able to agree to acceptable terms with the licensor or owner of those product candidates;
- our product candidates may not succeed in pre-clinical studies or clinical trials;

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- we may not succeed in formulation or process development;
- our product candidates 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 our product candidates obsolete or less attractive;
- product candidates that we develop may be covered by third parties' patents or other exclusive rights;
- product candidates that we develop may not allow us to leverage our expertise and our development and commercial infrastructure as currently expected;
- the market for a product candidate may change during our program so that such a product may become unreasonable to continue to develop;
- a product candidate may not be capable of being produced in commercial quantities at an acceptable cost, or at all; and
- a product candidate 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 our growth strategy may not deliver the anticipated results.

### **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.

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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. 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 seek 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

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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.

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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 former employees or consultants asserting an ownership right in our patents or patent applications, as a result of the work they performed on our behalf. 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 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 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,

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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 addition, we licensed KPL-045 from Novo Nordisk in August 2017 and the right to conduct research and development of KPL-404 from Primatope in September 2017. These current agreements and any future such agreements that we enter into impose a variety of obligations.

We are currently 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 all rights and remedies available to us, 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.

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

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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 oncostatin M receptor (other than for the development and commercialization of products that are the subject of the Biogen Agreement). This exclusivity obligation runs from the earlier of the eighth anniversary of the agreement or the first commercial sale of a product that is the subject of the agreement.

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

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

Given the vast number of patents in our field of technology, we cannot be certain that we do not infringe existing patents or that we will not infringe patents that may be granted in the future. Many companies have filed, and continue to file, patent applications related to immunomodulation. Some of these patent applications have already been allowed or issued, and others may issue in the future. For example, we are aware of third-party patents that contain claims potentially relevant to certain therapeutic uses of mavrimumab and KPL-716. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to mavrimumab 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

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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

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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.

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***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.

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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

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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

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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.

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***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,

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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 drug is approved by the FDA or similar regulatory authorities outside the United States. Moreover, eligibility for reimbursement does not imply that any drug 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 drugs 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.

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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. 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 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

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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 or they 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;

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- 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 a 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:

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- 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 services; 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 physician payments and other transfers of value and the ownership and investment interests of such 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.

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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

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- the inability to commercialize any product candidates that we may develop.

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

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

In the United States, 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, in March 2010, the Affordable Care Act was passed, which 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. It is uncertain the extent to which any such changes may impact our business or financial condition.

In addition, other legislative changes have been proposed and adopted in the United States since the Affordable Care Act was enacted. On August 2, 2011, 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. On January 2, 2013, the American Taxpayer Relief Act of 2012 was signed into law, which, 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 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. 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.

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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 healthcare 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;

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- 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

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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 Euros, 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.

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***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.

***We may acquire businesses, or products or product candidates, or form strategic alliances, in the future, and we may not realize the benefits of such acquisitions.***

We have acquired and in-licensed, and may acquire or in-license additional businesses or products, from other companies or create joint ventures with third parties that we believe will complement or augment our existing business. If we acquire businesses with promising markets or technologies, we may not be able to realize the benefit of acquiring such businesses if we are unable to successfully integrate them with our existing operations and company culture. We may encounter numerous difficulties in developing, manufacturing and marketing any new products resulting from a strategic alliance or acquisition that delay or prevent us from realizing their expected benefits or enhancing our business. We cannot assure you that, following any such acquisition or license, we will achieve the expected synergies to justify the transaction.

## Risks Related to 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. 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

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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. As of June 30, 2018, the holders of Class A common shares account for 25.4% of our aggregate voting power and the holders of Class B common shares account for the remaining 74.6% of our aggregate voting power. 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. 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.

As of June 30, 2018, our executive officers and certain other members of our senior management hold substantially all of our Class B common shares. 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 hold 69.7% of our voting power and have the ability to control the outcome of all matters submitted to our shareholders for approval.

However, this percentage may change depending on any conversion of Class A1 common shares, Class B1 common shares or Class B common shares. Each holder of 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 their ability to significantly influence or control matters submitted to our shareholders for approval.

As of June 30, 2018, entities managed by Baker Bros. Advisors LP hold 59.7% 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.

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

Our share price is likely to be volatile. The shares 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, holders of our Class A common shares may not be able to sell their Class A common shares at or above the price at which they purchased such shares. The market price for our Class A common shares may be influenced by many factors, including:

- the results of clinical trials for our product candidates;

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- 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 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 Pre-IPO 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 lapse, the market price of our Class A common shares could decline. As of June 30, 2018, we have outstanding a total of 15,754,771 Class A

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common shares, 12,995,954 Class A1 common shares, 4,638,855 Class B common shares and 16,057,618 Class B1 common shares assuming no exercise of options to purchase Class A common shares outstanding as of June 30, 2018. Of these shares, only 9,484,202 are freely tradable, without restriction, in the public market.

Substantially all of our shareholders prior to the IPO have entered into lock-up agreements pertaining to the IPO that restrict their ability to sell or transfer their common shares. The lock-up agreements will expire 180 days from May 23, 2018. After the lock-up agreements expire, up to an additional 39,962,996 Class A common shares 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). Approximately 27,551,086 of these Class A common shares will be held by our directors, executive officers and certain entities affiliated with our directors, and will, following the expiration of the lock-up, remain subject to certain limitations on sales made by affiliates pursuant to Rule 144 under the Securities Act of 1933, as amended, or the Securities Act. In addition, our Class A1 common shares, Class B common shares and Class B1 common shares automatically convert into Class A common shares upon transfer to non-affiliates. As a result, up to 28,449,865 of our Class A common shares may be issued upon such transfers. The representatives of the underwriters for the IPO may, in their sole discretion, permit our officers, directors and other shareholders who are subject to these lock-up agreements to sell shares prior to the expiration of the lock-up.

In addition, as of June 30, 2018 4,805,037 of our Class A common shares that are subject to outstanding options under our equity incentive plans may become eligible for sale in the public market to the extent permitted by the provisions of various vesting schedules, the lock-up agreements and Rule 144 and Rule 701 under the Securities Act. If 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.

The holders of approximately 35,670,093 of our 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) are entitled to rights with respect to the registration of their shares under the Securities Act, subject 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.

***Future sales and issuances of our common shares or rights to purchase common shares, including pursuant to our equity incentive plans, could result in 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 A1 common shares, Class B 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 are an “emerging growth company,” and the reduced disclosure requirements applicable to emerging growth companies may make our Class A common shares less attractive to investors.***

We are an “emerging growth company,” as defined in the Jumpstart Our Business Startups Act, or JOBS Act. 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 Class A 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:

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- 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 and shareholder approval of any golden parachute payments not previously approved.

We may choose to take advantage of some, but not all, of the available exemptions. 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.

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 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, and particularly after we are no longer an “emerging growth 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. 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 of the Sarbanes-Oxley Act of 2002, or 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.

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***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<sup>2</sup>/<sub>3</sub>% of the voting power of our voting shares for certain “business combination” transactions that have not been approved by our board of directors;
- our multi-class common share structure, which provides our holders of Class B common shares with the ability to significantly influence the outcome of matters requiring shareholder approval, even if they own less than a majority of our outstanding Class A common shares, Class A1 common shares, Class B 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.

***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 the sole source of gain for our shareholders for the foreseeable future.

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

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

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

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***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 circumstances in which shareholder derivative actions may be available under Bermuda law are substantially more proscribed and less clear than they would be to shareholders of U.S. corporations. The Bermuda courts, however, would ordinarily be expected to permit a shareholder to commence an action in the name of a company to remedy a wrong to the company where the act complained of is alleged to be beyond the corporate power of the company or illegal, or would result in the

violation of the company's memorandum of association or bye-laws. Furthermore, consideration would be given by a Bermuda court to acts that are alleged to constitute a fraud against the minority shareholders or, for instance, where an act requires the approval of a greater percentage of the company's shareholders than those who actually approved it.

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

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

Common shares may be offered or sold in Bermuda only in compliance with the provisions of the Companies Act and the Bermuda Investment Business Act 2003, as amended, which regulates the sale of securities in Bermuda. In addition, the Bermuda Monetary Authority must approve all issues and transfers of shares of a Bermuda exempted company. However, the Bermuda Monetary Authority has, pursuant to its statement of June 1,

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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;

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- 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 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 Internal Revenue 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 we are classified as a PFIC for any taxable year during which a U.S. Holder holds our common shares, certain adverse U.S. federal income tax consequences could apply to such U.S. Holder, including (i) the treatment of all or a portion of any gain on disposition as ordinary income, (ii) the application of a deferred interest charge on such gain and the receipt of certain dividends and (iii) the obligation to comply with certain reporting requirements.

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

We believe we 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” and investments in U.S. property by controlled foreign corporations,

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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, limiting interest deductions, adopting elements of a territorial tax system and introducing certain anti-base erosion provisions. We continue to examine the impact this tax reform legislation may have on our business. The effect of the Tax Reform Act on our business, whether adverse or favorable, is uncertain, and may not become evident for some period of time. U.S. Holders should consult with their legal and tax advisors regarding any such legislation and the potential tax consequences of investing in our common shares.

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

**Use of Proceeds**

On May 29, 2018, we issued and sold 8,477,777 Class A common shares to the underwriters of our IPO and on June 22, 2018, we issued and sold an additional 1,006,425 Class A common shares pursuant to the exercise by the underwriters of their over-allotment option to purchase additional shares. Our Class A common shares were sold at a price to the public of \$18.00 per share.

The offer and sale of all of the shares in our IPO inclusive of the underwriters’ over-allotment option were registered under the Securities Act pursuant to a registration statement on Form S-1 (File No. 333-224466), which was declared effective by the Securities and Exchange Commission on May 23, 2018, and a

registration statement on Form S-1 to register additional shares (File No. 333-225159), which was automatically effective upon filing with the Securities and Exchange Commission on May 23, 2018. The IPO commenced on May 14, 2018 and terminated upon the closing of the sale of shares to the underwriters on June 22, 2018 under the underwriters' over-allotment option. Goldman Sachs & Co. LLC and J.P. Morgan Securities LLC acted as joint book-running managers for the IPO and JMP Securities LLC and Wedbush Securities Inc. acted as co-managers of the IPO.

We received aggregate gross proceeds from the IPO inclusive of the underwriters' over-allotment option of approximately \$170.7 million and aggregate net proceeds of approximately \$155.5 million after deducting underwriting discounts and commissions and other offering expenses. No payments for such expenses were made directly or indirectly to (i) any of our officers or directors or their associates, (ii) any persons owning 10% or more of any class of our equity securities or (iii) any of our affiliates.

There has been no material change in our planned use of the net proceeds from the IPO as described in our final prospectus filed pursuant to Rule 424(b) (4) under the Securities Act with the Securities and Exchange Commission on May 24, 2018.

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**Item 3. Defaults Upon Senior Securities.**

None.

**Item 4. Mine Safety Disclosures.**

None.

**Item 5. Other Information.**

None.

**Item 6. Exhibits**

Exhibit Number	Exhibit Description	Incorporated by Reference				Filed/ Furnished Herewith
		Form	File No.	Exhibit	Filing Date	
3.1	<a href="#">Amended and Restated Bye-Laws of Kiniksa Pharmaceuticals, Ltd.</a>	8-K	001-38492	3.1	5/29/18	
4.1	<a href="#">Specimen Share Certificate evidencing the Class A common shares</a>	S-1/A	333-224488	4.1	5/14/18	
4.2	<a href="#">Second Amended and Restated Investors' Rights Agreement, dated as of February 9, 2018</a>	S-1	333-224488	4.2	4/27/18	
10.1	<a href="#">2018 Incentive Award Plan</a>					*
10.2	<a href="#">Form of Share Option Grant Notice and Share Option Agreement under the 2018 Incentive Award Plan</a>					*
10.3	<a href="#">Form of Restricted Share Grant Notice and Restricted Share Agreement under the 2018 Incentive Award Plan</a>					*
10.4	<a href="#">Form of Restricted Share Unit Grant Notice and Restricted Share Unit Agreement under the 2018 Incentive Award Plan</a>					*
10.5	<a href="#">2018 Employee Share Purchase Plan</a>					*
10.6	<a href="#">Offering Document under the 2018 Employee Share Purchase Plan</a>					*

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10.7	<a href="#">Amended and Restated Employment Agreement, dated as of May 29, 2018, by and between Kiniksa Pharmaceuticals Corp. and Sanj K. Patel</a>					*
10.8	<a href="#">Amended and Restated Employment Agreement, dated as of May 29, 2018, by and between Kiniksa Pharmaceuticals Corp. and Stephen Mahoney</a>					*
10.9	<a href="#">Amended and Restated Employment Agreement, dated as of May 29, 2018, by and between Kiniksa Pharmaceuticals Corp. and John F. Paolini</a>					*

10.10	<a href="#">First and Second Amendment to Sublease Agreement, dated as of June 26, 2018 and July 17, 2018, respectively, by and between Kiniksa Pharmaceuticals Corp. and Shire Human Genetic Pharmaceuticals Corp.</a>	*
31.1	<a href="#">Rule 13a-14(a) / 15d-14(a) Certification of Chief Executive Officer</a>	*
31.2	<a href="#">Rule 13a-14(a) / 15d-14(a) Certification of Chief Financial Officer</a>	*
32.1	<a href="#">Section 1350 Certification of Chief Executive Officer</a>	**
32.2	<a href="#">Section 1350 Certification of Chief Financial Officer</a>	**
101.INS	XBRL Instance Document	***
101.SCH	XBRL Taxonomy Extension Schema Document	***
101.CAL	XBRL Taxonomy Extension Calculation Linkbase Document	***
101.DEF	XBRL Extension Definition Linkbase Document	***
101.LAB	XBRL Taxonomy Label Linkbase Document	***
101.PRE	XBRL Taxonomy Extension Presentation Linkbase Document	***

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

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**SIGNATURES**

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

**KINIKSA PHARMACEUTICALS, LTD.**

Date: August 6, 2018

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

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

**ARTICLE I.  
PURPOSE**

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

**ARTICLE II.  
ELIGIBILITY**

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

**ARTICLE III.  
ADMINISTRATION AND DELEGATION**

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

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

**ARTICLE IV.  
SHARES AVAILABLE FOR AWARDS**

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

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

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

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

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

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

5.1 General. The Administrator may grant Options or Share Appreciation Rights to Service Providers subject to the limitations in the Plan, including any limitations in the Plan that apply to Incentive Stock Options. The Administrator will determine the number of Shares covered by each Option

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

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

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

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

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

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

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

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

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

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

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

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

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

## ARTICLE VI. RESTRICTED SHARES; RESTRICTED SHARE UNITS

6.1 General. The Administrator may grant Restricted Shares, or the right to purchase Restricted Shares, to any Service Provider, subject to the Company's right to repurchase all or part of such shares at their issue price or other stated or formula price from the Participant (or to require forfeiture of

such shares) if conditions the Administrator specifies in the Award Agreement are not satisfied before the end of the applicable restriction period or periods that the Administrator establishes for such Award and subject to Applicable Laws. In addition, the Administrator may grant to Service Providers Restricted Share Units, which may be subject to vesting and forfeiture conditions during the applicable restriction period or periods, as set forth in an Award Agreement. The Administrator will determine and set forth in the Award Agreement the terms and conditions for each Restricted Share and Restricted Share Unit Award, subject to the conditions and limitations contained in the Plan.

## 6.2 Restricted Shares.

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

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

## 6.3 Restricted Share Units.

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

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

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

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

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

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

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

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

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

(a) To provide for the cancellation of any such Award in exchange for either an amount of cash or other property with a value equal to the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participant's rights under

the vested portion of such Award, as applicable; provided that, if the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participant's rights, in any case, is equal to or less than zero, then the Award may be terminated without payment;

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

## ARTICLE X. MISCELLANEOUS

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

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

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

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

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

10.6 Section 409A.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

## ARTICLE XI. DEFINITIONS

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

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

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

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

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

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

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

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

determination of the Administrator as to the existence of Cause will be conclusive on the Participant and the Company.

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

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

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

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

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

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

11.10 **“Common Shares”** means the Class A Common Shares of the Company of par value US\$0.000273235 each.

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

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

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

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

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

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

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

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

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

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

will determine the Fair Market Value in its discretion. Notwithstanding the foregoing, with respect to any Award granted on the pricing date of the Company's initial public offering, the Fair Market Value shall

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

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

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

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

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

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

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

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

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

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

11.29 "**Plan**" means this 2018 Incentive Award Plan.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

**SHARE OPTION GRANT NOTICE**

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

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

**Participant:**

**Grant Date:**

**Exercise Price per Share:**

**Shares Subject to the Option:**

**Final Expiration Date:**

**Vesting Commencement Date:**

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

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

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

**KINIKSA PHARMACEUTICALS, LTD.**

**PARTICIPANT**

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

Name: \_\_\_\_\_

[Participant Name]

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

**Exhibit A**

**SHARE OPTION AGREEMENT**

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

**ARTICLE I.  
GENERAL**

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

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

**ARTICLE II.  
PERIOD OF EXERCISABILITY**

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

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

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

(a) The final expiration date in the Grant Notice;

(b) Except as the Administrator may otherwise approve, the expiration of three (3) months from the date of Participant's Termination of Service, unless Participant's Termination of Service is for Cause or by reason of Participant's death or Disability;

(c) Except as the Administrator may otherwise approve, the expiration of one (1) year from the date of Participant's Termination of Service by reason of Participant's death or Disability; and

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

### ARTICLE III. EXERCISE OF OPTION

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

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

3.3 Tax Withholding.

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

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

### ARTICLE IV. OTHER PROVISIONS

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

**RESTRICTED SHARE GRANT NOTICE**

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

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

**Participant:**

**Grant Date:**

**Number of Restricted Shares:**

**Vesting Commencement Date:**

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

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

**KINIKSA PHARMACEUTICALS, LTD.**

**PARTICIPANT**

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

\_\_\_\_\_

Name: \_\_\_\_\_

[Participant Name]

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

**Exhibit A**

**RESTRICTED SHARE AGREEMENT**

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

**ARTICLE I.  
GENERAL**

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

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

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

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

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

2.3 **Escrow.**

(a) Unvested Shares in certificated form will be held by the Company or its authorized representatives until (i) they are forfeited, (ii) they become Vested Shares or (iii) this Agreement is no longer in effect. By accepting this Award, Participant appoints the Company and its authorized

representatives as Participant's attorney(s)-in-fact to take all actions necessary to effect cancellation of forfeited Unvested Shares (and Retained Distributions (as defined below), if any, paid on such forfeited Unvested Shares) to the Company as may be required pursuant to the Plan or this Agreement and to execute such representations or other documents or assurances as the Company or such representatives deem necessary or advisable in connection with any such transfer. The Company, or its authorized representative, will not be liable for any good faith act or omission with respect to the holding in escrow or transfer of the Restricted Shares.

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

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

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

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

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

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

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

3.3 **Tax Withholding.**

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

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

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### **ARTICLE IV. RESTRICTIVE LEGENDS AND TRANSFERABILITY**

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

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

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

### **ARTICLE V. OTHER PROVISIONS**

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

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

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

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

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

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

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

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

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

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

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

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

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

**RESTRICTED SHARE UNIT GRANT NOTICE**

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

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

**Participant:**

**Grant Date:**

**Number of RSUs:**

**Vesting Commencement Date:**

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

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

**KINIKSA PHARMACEUTICALS, LTD.**

**PARTICIPANT**

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

Name: \_\_\_\_\_

[Participant Name]

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

**Exhibit A**

**RESTRICTED SHARE UNIT AGREEMENT**

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

**ARTICLE I.  
GENERAL**

**1.1 Award of RSUs and Dividend Equivalents.**

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

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

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

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

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

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

will vest or be forfeited, as applicable, upon the vesting or forfeiture of the RSU with respect to which the Dividend Equivalent (including the Dividend Equivalent Account) relates.

## 2.2 Settlement.

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

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

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

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

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

## 3.2 Tax Withholding.

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

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

## **ARTICLE IV. OTHER PROVISIONS**

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

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

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

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

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

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

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

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

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

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

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

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

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**KINIKSA PHARMACEUTICALS, LTD.  
2018 EMPLOYEE SHARE PURCHASE PLAN**

**ARTICLE I.  
PURPOSE**

The purposes of this Kiniksa Pharmaceuticals, Ltd. 2018 Employee Share Purchase Plan (as it may be amended or restated from time to time, the “*Plan*”) are to assist Eligible Employees of Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company (the “*Company*”), and its Designated Subsidiaries in acquiring an ownership interest in the Company pursuant to a plan which is intended to qualify as an “employee stock purchase plan” within the meaning of Section 423(b) of the Code, and to help Eligible Employees provide for their future security and to encourage them to remain in the employment of the Company and its Designated Subsidiaries.

**ARTICLE II.  
DEFINITIONS AND CONSTRUCTION**

Wherever the following terms are used in the Plan they shall have the meanings specified below, unless the context clearly indicates otherwise. The singular pronoun shall include the plural where the context so indicates. Masculine, feminine and neuter pronouns are used interchangeably and each comprehends the others.

2.1 “*Administrator*” shall mean the entity that conducts the general administration of the Plan as provided in Article XI. The term “Administrator” shall refer to the Committee unless the Board has assumed the authority for administration of the Plan as provided in Article XI.

2.2 “*Applicable Law*” shall mean the requirements relating to the administration of equity incentive plans under U.S. federal and state securities, tax and other applicable laws, rules and regulations, the applicable rules of any stock exchange or quotation system on which the Common Shares are listed or quoted and the applicable laws and rules of any foreign country or other jurisdiction where rights under this Plan are granted, including without limitation, the laws of Bermuda.

2.3 “*Board*” shall mean the Board of Directors of the Company.

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

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outstanding voting shares as a result of a repurchase or other acquisition of voting shares by the Company reducing the number of shares outstanding, provided that if a Change in Control would occur (but for the operating of this sentence) as a result of the acquisition of voting shares by the Company, and after such share acquisition, the Subject Person becomes the owner of any additional voting shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

The Administrator shall have full and final authority, which shall be exercised in its discretion, to determine conclusively whether a Change in Control has occurred pursuant to the above definition, the date of the occurrence of such Change in Control and any incidental matters relating thereto.

2.5 “*Code*” shall mean the Internal Revenue Code of 1986, as amended, and the regulations issued thereunder.

2.6 “*Common Shares*” shall mean the Class A Common Shares of the Company of par value US\$0.000273235 each.

2.7 “*Company*” shall mean of Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company, or any successor.

2.8 “*Compensation*” of an Eligible Employee shall mean the gross base compensation received by such Eligible Employee as compensation for services to the Company or any Designated Subsidiary, including overtime payments and excluding sales commissions, incentive compensation, bonuses, expense reimbursements, fringe benefits and other special payments.

2.9 “*Designated Subsidiary*” shall mean any Subsidiary designated by the Administrator in accordance with Section 11.3(b).

2.10 “*Effective Date*” shall mean the day prior to the Public Trading Date.

2.11 “*Eligible Employee*” shall mean an Employee: (a) who does not, immediately after any rights under this Plan are granted, own (directly or through attribution) shares possessing 5% or more of the total combined voting power or value of all classes of Common Shares and other shares of the Company, a Parent or a Subsidiary (as determined under Section 423(b)(3) of the Code); (b) whose customary employment is for more than twenty hours per week; and (c) whose customary employment is for more than five months in any calendar year. For purposes of the foregoing, the rules of Section 424(d) of the Code with regard to the attribution of share ownership shall apply in determining the share ownership of an individual, and shares that an Employee may purchase under outstanding options shall be treated as shares owned by the Employee; provided, however, that the Administrator may provide in an Offering Document that an Employee shall not be eligible to participate in an Offering Period if: (i) such Employee is a highly compensated employee within the

meaning of Section 423(b)(4)(D) of the Code; and/or (ii) such Employee has not met a service requirement designated by the Administrator pursuant to Section 423(b)(4)(A) of the Code (which service requirement may not exceed two years), and/or (iii) such Employee is a citizen or resident of a foreign jurisdiction and the grant of a right to purchase Common Shares under the Plan to such Employee would be prohibited under the laws of such foreign jurisdiction or the grant of a right to purchase Common Shares under the Plan to such Employee in compliance with the laws of such foreign jurisdiction would cause the Plan to violate the requirements of Section 423 of the Code, as determined by the Administrator in its sole discretion; provided, further, that any exclusion in clauses (i), (ii) or (iii) shall

be applied in an identical manner under each Offering Period to all Employees, in accordance with Treasury Regulation Section 1.423-2(e).

2.12 “**Employee**” shall mean any officer or other employee (as defined in accordance with Section 3401(c) of the Code) of the Company or any Designated Subsidiary. “Employee” shall not include any director of the Company or a Designated Subsidiary who does not render services to the Company or a Designated Subsidiary as an employee within the meaning of Section 3401(c) of the Code. For purposes of the Plan, the employment relationship shall be treated as continuing intact while the individual is on sick leave or other leave of absence approved by the Company or Designated Subsidiary and meeting the requirements of Treasury Regulation Section 1.421-1(h)(2). Where the period of leave exceeds three (3) months and the individual’s right to reemployment is not guaranteed either by statute or by contract, the employment relationship shall be deemed to have terminated on the first day immediately following such three (3)-month period.

2.13 “**Enrollment Date**” shall mean the first Trading Day of each Offering Period.

2.14 “**Exchange Act**” shall mean the Securities Exchange Act of 1934, as amended.

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

2.16 “**Offering Document**” shall have the meaning given to such term in Section 4.1.

2.17 “**Offering Period**” shall have the meaning given to such term in Section 4.1.

2.18 “**Parent**” shall mean any corporation, other than the Company, in an unbroken chain of corporations ending with the Company if, at the time of the determination, each of the corporations other than the Company owns shares possessing 50% or more of the total combined voting power of all classes of shares in one of the other corporations in such chain.

2.19 “**Participant**” shall mean any Eligible Employee who has executed a subscription agreement and been granted rights to purchase Common Shares pursuant to the Plan.

2.20 “**Plan**” shall mean this 2018 Employee Share Purchase Plan.

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

2.22 “**Purchase Date**” shall mean the last Trading Day of each Offering Period.

2.23 “**Purchase Price**” shall mean the purchase price designated by the Administrator in the applicable Offering Document (which purchase price shall not be less than 85% of the Fair Market Value of a Share on the Enrollment Date or on the Purchase Date, whichever is lower); provided, however, that, in the event no purchase price is designated by the Administrator in the applicable Offering Document, the purchase price for the Offering Periods covered by such Offering Document shall be 85% of the Fair Market Value of a Share on the Enrollment Date or on the Purchase Date, whichever is lower; provided, further, that the Purchase Price may be adjusted by the Administrator pursuant to Article VIII and shall not be less than the par value of a Share.

2.24 “**Securities Act**” shall mean the Securities Act of 1933, as amended.

2.25 “**Share**” shall mean a Common Share.

2.26 “**Subject Person**” shall have the meaning given to such term in Section 2.4.

2.27 “**Subsidiary**” shall mean any corporation, other than the Company, in an unbroken chain of corporations beginning with the Company if, at the time of the determination, each of the corporations other than the last corporation in an unbroken chain owns shares possessing 50% or more of the total combined voting power of all classes of shares in one of the other corporations in such chain; provided, however, that a limited liability company or partnership may be treated as a Subsidiary to the extent either (a) such entity is treated as a disregarded entity under Treasury Regulation Section 301.7701-3(a) by reason of the Company or any other Subsidiary that is a corporation being the sole owner of such entity, or (b) such entity elects to be classified as a corporation under Treasury Regulation Section 301.7701-3(a) and such entity would otherwise qualify as a Subsidiary.

**ARTICLE III.  
SHARES SUBJECT TO THE PLAN**

3.1 Number of Shares. Subject to Article VIII, the aggregate number of Shares that may be issued pursuant to rights granted under the Plan shall be 670,000 Shares. In addition to the foregoing, subject to Article VIII, on the first day of each calendar year beginning on January 1, 2019 and ending on and including January 1, 2028, the number of Shares available for issuance under the Plan shall be increased by that number of Shares equal to the lesser of (a) 1% of the Shares outstanding (on an as-converted basis) on the final day of the immediately preceding calendar year and (b) such smaller number of Shares as determined by the Board. If any right granted under the Plan shall for any reason terminate without having been exercised, the Common Shares not purchased under such right shall again become available for issuance under the Plan. Notwithstanding anything in this Section 3.1 to the contrary, the number of Shares that may be issued or transferred pursuant to the rights granted under the Plan shall not exceed an aggregate of 6,420,000 Shares, subject to Article VIII.

3.2 Shares Distributed. Any Common Shares distributed pursuant to the Plan may consist, in whole or in part, of authorized and unissued Common Shares, Common Shares held in treasury or Common Shares purchased on the open market.

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**ARTICLE IV.  
OFFERING PERIODS; OFFERING DOCUMENTS; PURCHASE DATES**

4.1 Offering Periods. The Administrator may from time to time grant or provide for the grant of rights to purchase Common Shares under the Plan to Eligible Employees during one or more periods (each, an “*Offering Period*”) selected by the Administrator. The terms and conditions applicable to each Offering Period shall be set forth in an “*Offering Document*” adopted by the Administrator, which Offering Document shall be in such form and shall contain such terms and conditions as the Administrator shall deem appropriate and shall be incorporated by reference into and made part of the Plan and shall be attached hereto as part of the Plan. The provisions of separate Offering Periods under the Plan need not be identical.

4.2 Offering Documents. Each Offering Document with respect to an Offering Period shall specify (through incorporation of the provisions of this Plan by reference or otherwise):

(a) the length of the Offering Period, which period shall not exceed twenty-seven months;

(b) the maximum number of Shares that may be purchased by any Eligible Employee during such Offering Period, which, in the absence of a contrary designation by the Administrator, shall be 25,000 Shares; and

(c) such other provisions as the Administrator determines are appropriate, subject to the Plan.

**ARTICLE V.  
ELIGIBILITY AND PARTICIPATION**

5.1 Eligibility. Any Eligible Employee who shall be employed by the Company or a Designated Subsidiary on a given Enrollment Date for an Offering Period shall be eligible to participate in the Plan during such Offering Period, subject to the requirements of this Article V and the limitations imposed by Section 423(b) of the Code.

5.2 Enrollment in Plan.

(a) Except as otherwise set forth in an Offering Document or determined by the Administrator, an Eligible Employee may become a Participant in the Plan for an Offering Period by delivering a subscription agreement to the Company by such time prior to the Enrollment Date for such Offering Period (or such other date specified in the Offering Document) designated by the Administrator and in such form as the Company provides.

(b) Each subscription agreement shall designate a whole percentage of such Eligible Employee’s Compensation to be withheld by the Company or the Designated Subsidiary employing such Eligible Employee on each payday during the Offering Period as payroll deductions under the Plan. The percentage of Compensation designated by an Eligible Employee may not be less than 1% and may not be more than the maximum percentage specified by the Administrator in the applicable Offering Document (which percentage shall be 25% in the absence of any such designation) as payroll deductions. The payroll deductions made for each Participant shall be credited to an account for such Participant under the Plan and shall be deposited with the general funds of the Company.

(c) A Participant may increase or decrease the percentage of Compensation designated in his or her subscription agreement, subject to the limits of this Section 5.2, or may suspend his or her payroll deductions, at any time during an Offering Period; provided, however, that the Administrator may limit the number of changes a Participant may make to his or her payroll deduction

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elections during each Offering Period in the applicable Offering Document (and in the absence of any specific designation by the Administrator, a Participant shall be allowed one change to his or her payroll deduction elections during each Offering Period). Any such change or suspension of payroll deductions shall be effective with the first full payroll period following five business days after the Company’s receipt of the new subscription agreement (or such shorter or longer period as may be specified by the Administrator in the applicable Offering Document). In the event a Participant suspends his or her payroll deductions, such Participant’s cumulative payroll deductions prior to the suspension shall remain in his or her account and shall be applied to the purchase of Shares on the next occurring Purchase Date and shall not be paid to such Participant unless he or she withdraws from participation in the Plan pursuant to Article VII.

(d) Except as otherwise set forth in an Offering Document or determined by the Administrator, a Participant may participate in the Plan only by means of payroll deduction and may not make contributions by lump sum payment for any Offering Period.

5.3 **Payroll Deductions.** Except as otherwise provided in the applicable Offering Document, payroll deductions for a Participant shall commence on the first payroll following the Enrollment Date and shall end on the last payroll in the Offering Period to which the Participant's authorization is applicable, unless sooner terminated by the Participant as provided in Article VII or suspended by the Participant or the Administrator as provided in Section 5.2 and Section 5.6, respectively.

5.4 **Effect of Enrollment.** A Participant's completion of a subscription agreement will enroll such Participant in the Plan for each subsequent Offering Period on the terms contained therein until the Participant either submits a new subscription agreement, withdraws from participation under the Plan as provided in Article VII or otherwise becomes ineligible to participate in the Plan.

5.5 **Limitation on Purchase of Common Shares.** An Eligible Employee may be granted rights under the Plan only if such rights, together with any other rights granted to such Eligible Employee under "employee stock purchase plans" of the Company, any Parent or any Subsidiary, as specified by Section 423(b)(8) of the Code, do not permit such employee's rights to purchase shares of the Company or any Parent or Subsidiary to accrue at a rate that exceeds \$25,000 of the fair market value of such shares (determined as of the first day of the Offering Period during which such rights are granted) for each calendar year in which such rights are outstanding at any time. This limitation shall be applied in accordance with Section 423(b)(8) of the Code.

5.6 **Decrease or Suspension of Payroll Deductions.** Notwithstanding the foregoing, to the extent necessary to comply with Section 423(b)(8) of the Code and Section 5.5 or the other limitations set forth in this Plan, a Participant's payroll deductions may be suspended by the Administrator at any time during an Offering Period. The balance of the amount credited to the account of each Participant that has not been applied to the purchase of Shares by reason of Section 423(b)(8) of the Code, Section 5.5 or the other limitations set forth in this Plan shall be paid to such Participant in one lump sum in cash as soon as reasonably practicable after the Purchase Date.

5.7 **Non-U.S. Employees.** In order to facilitate participation in the Plan, the Administrator may provide for such special terms applicable to Participants who are citizens or residents of a country other than the United States, or who are employed by a Designated Subsidiary outside of the United States, as the Administrator may consider necessary or appropriate to accommodate differences in local law, tax policy or custom. Such special terms may not be more favorable than the terms of rights granted under the Plan to Eligible Employees who are residents of the United States. Moreover, the Administrator may approve such supplements to, or amendments, restatements or alternative versions of, this Plan as it may consider necessary or appropriate for such purposes without thereby affecting the

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terms of this Plan as in effect for any other purpose. No such special terms, supplements, amendments or restatements shall include any provisions that are inconsistent with the terms of this Plan as then in effect unless this Plan could have been amended to eliminate such inconsistency without further approval by the shareholders of the Company.

5.8 **Leave of Absence.** During leaves of absence approved by the Company meeting the requirements of Treasury Regulation Section 1.421-1(h)(2) under the Code, a Participant may continue participation in the Plan by making cash payments to the Company on his or her normal payday equal to his or her authorized payroll deduction.

## **ARTICLE VI. GRANT AND EXERCISE OF RIGHTS**

6.1 **Grant of Rights.** On the Enrollment Date of each Offering Period, each Eligible Employee participating in such Offering Period shall be granted a right to purchase the maximum number of Shares specified under Section 4.2, subject to the limits in Section 5.5, and shall have the right to buy, on each Purchase Date during such Offering Period (at the applicable Purchase Price), such number of whole Shares as is determined by dividing (a) such Participant's payroll deductions accumulated prior to such Purchase Date and retained in the Participant's account as of the Purchase Date, by (b) the applicable Purchase Price (rounded down to the nearest Share). The right shall expire on the last day of the Offering Period.

6.2 **Exercise of Rights.** On each Purchase Date, each Participant's accumulated payroll deductions and any other additional payments specifically provided for in the applicable Offering Document will be applied to the purchase of whole Shares, up to the maximum number of Shares permitted pursuant to the terms of the Plan and the applicable Offering Document, at the Purchase Price. No fractional Shares shall be issued upon the exercise of rights granted under the Plan, unless the Offering Document specifically provides otherwise. Any cash in lieu of fractional Shares remaining after the purchase of whole Shares upon exercise of a purchase right will be credited to a Participant's account and carried forward and applied toward the purchase of whole Shares for the next following Offering Period. Shares issued pursuant to the Plan may be evidenced in such manner as the Administrator may determine and may be issued in certificated form or issued pursuant to book-entry procedures.

6.3 **Pro Rata Allocation of Shares.** If the Administrator determines that, on a given Purchase Date, the number of Shares with respect to which rights are to be exercised may exceed (a) the number of Shares that were available for issuance under the Plan on the Enrollment Date of the applicable Offering Period, or (b) the number of Shares available for issuance under the Plan on such Purchase Date, the Administrator may in its sole discretion provide that the Company shall make a pro rata allocation of the Shares available for purchase on such Enrollment Date or Purchase Date, as applicable, in as uniform a manner as shall be practicable and as it shall determine in its sole discretion to be equitable among all Participants for whom rights to purchase Common Shares are to be exercised pursuant to this Article VI on such Purchase Date, and shall either (i) continue all Offering Periods then in effect, or (ii) terminate any or all Offering Periods then in effect pursuant to Article IX. The Company may make pro rata allocation of the Shares available on the Enrollment Date of any applicable Offering Period pursuant to the preceding sentence, notwithstanding any authorization of additional Shares for issuance under the Plan by the Company's shareholders subsequent to such Enrollment Date. The balance of the amount credited to the account of each Participant that has not been applied to the purchase of Shares shall be paid to such Participant in one lump sum in cash as soon as reasonably practicable after the Purchase Date.

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6.4 Withholding. At the time a Participant's rights under the Plan are exercised, in whole or in part, or at the time some or all of the Common Shares issued under the Plan is disposed of, the Participant must make adequate provision for the Company's federal, state, or other tax withholding obligations, if any, that arise upon the exercise of the right or the disposition of the Common Shares. At any time, the Company may, but shall not be obligated to, withhold from the Participant's compensation the amount necessary for the Company to meet applicable withholding obligations, including any withholding required to make available to the Company any tax deductions or benefits attributable to sale or early disposition of Common Shares by the Participant.

6.5 Conditions to Issuance of Common Shares. The Company shall not be required to issue or deliver any certificate or certificates for, or make any book entries evidencing, Shares purchased upon the exercise of rights under the Plan prior to fulfillment of all of the following conditions:

(a) The admission of such Shares to listing on all stock exchanges, if any, on which the Common Shares are then listed;

(b) The completion of any registration or other qualification of such Shares under any state or federal law or under the rulings or regulations of the Securities and Exchange Commission or any other governmental regulatory body, that the Administrator shall, in its absolute discretion, deem necessary or advisable;

(c) The obtaining of any approval or other clearance from any state or federal governmental agency that the Administrator shall, in its absolute discretion, determine to be necessary or advisable;

(d) The payment to the Company of all amounts that it is required to withhold under federal, state or local law upon exercise of the rights, if any; and

(e) The lapse of such reasonable period of time following the exercise of the rights as the Administrator may from time to time establish for reasons of administrative convenience.

## ARTICLE VII.

### WITHDRAWAL; CESSATION OF ELIGIBILITY

7.1 Withdrawal. A Participant may withdraw all but not less than all of the payroll deductions credited to his or her account and not yet used to exercise his or her rights under the Plan at any time by giving written notice to the Company in a form acceptable to the Company no later than one week prior to the end of the Offering Period. All of the Participant's payroll deductions credited to his or her account during an Offering Period shall be paid to such Participant as soon as reasonably practicable after receipt of notice of withdrawal and such Participant's rights for the Offering Period shall be automatically terminated, and no further payroll deductions for the purchase of Shares shall be made for such Offering Period. If a Participant withdraws from an Offering Period, payroll deductions shall not resume at the beginning of the next Offering Period unless the Participant timely delivers to the Company a new subscription agreement.

7.2 Future Participation. A Participant's withdrawal from an Offering Period shall not have any effect upon his or her eligibility to participate in any similar plan that may hereafter be adopted by the Company or a Designated Subsidiary or in subsequent Offering Periods that commence after the termination of the Offering Period from which the Participant withdraws.

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7.3 Cessation of Eligibility. Upon a Participant's ceasing to be an Eligible Employee for any reason, he or she shall be deemed to have elected to withdraw from the Plan pursuant to this Article VII and the payroll deductions credited to such Participant's account during the Offering Period shall be paid to such Participant or, in the case of his or her death, to the person or persons entitled thereto under Section 12.4, as soon as reasonably practicable, and such Participant's rights for the Offering Period shall be automatically terminated.

## ARTICLE VIII.

### ADJUSTMENTS UPON CHANGES IN SHARES

8.1 Changes in Capitalization. Subject to Section 8.3, in the event that the Administrator determines that any dividend or other distribution (whether in the form of cash, Common Shares, other securities, or other property), Change in Control, reorganization, merger, amalgamation, consolidation, combination, repurchase, recapitalization, liquidation, dissolution, or sale, transfer, exchange or other disposition of all or substantially all of the assets of the Company, or sale or exchange of Common Shares or other securities of the Company, issuance of warrants or other rights to purchase Common Shares or other securities of the Company, or other similar corporate transaction or event, as determined by the Administrator, affects the Common Shares such that an adjustment is determined by the Administrator to be appropriate in order to prevent dilution or enlargement of the benefits or potential benefits intended by the Company to be made available under the Plan or with respect to any outstanding purchase rights under the Plan, the Administrator shall make equitable adjustments, if any, to reflect such change with respect to (a) the aggregate number and type of Shares (or other securities or property) that may be issued under the Plan (including, but not limited to, adjustments of the limitations in Section 3.1 and the limitations established in each Offering Document pursuant to Section 4.2 on the maximum number of Shares that may be purchased); (b) the class(es) and number of Shares and price per Share subject to outstanding rights; and (c) the Purchase Price with respect to any outstanding rights.

8.2 Other Adjustments. Subject to Section 8.3, in the event of any transaction or event described in Section 8.1 or any unusual or nonrecurring transactions or events affecting the Company, any affiliate of the Company, or the financial statements of the Company or any affiliate (including without limitation any Change in Control), or of changes in Applicable Law or accounting principles, the Administrator, in its discretion, and on such terms and conditions as it deems appropriate, is hereby authorized to take any one or more of the following actions whenever the Administrator determines that such action is appropriate in order to prevent the dilution or enlargement of the benefits or potential benefits intended to be made available under the Plan or with respect to any right under the Plan, to facilitate such transactions or events or to give effect to such changes in laws, regulations or principles:

(a) To provide for either (i) termination of any outstanding right in exchange for an amount of cash, if any, equal to the amount that would have been obtained upon the exercise of such right had such right been currently exercisable or (ii) the replacement of such outstanding right with other rights or property selected by the Administrator in its sole discretion;

(b) To provide that the outstanding rights under the Plan shall be assumed by the successor or survivor corporation, or a parent or subsidiary thereof, or shall be substituted for by similar rights covering the shares of the successor or survivor corporation, or a parent or subsidiary thereof, with appropriate adjustments as to the number and kind of shares and prices;

(c) To make adjustments in the number and type of Shares (or other securities or property) subject to outstanding rights under the Plan and/or in the terms and conditions of outstanding rights and rights that may be granted in the future;

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(d) To provide that Participants' accumulated payroll deductions may be used to purchase Common Shares prior to the next occurring Purchase Date on such date as the Administrator determines in its sole discretion and the Participants' rights under the ongoing Offering Period(s) shall be terminated; and

(e) To provide that all outstanding rights shall terminate without being exercised.

8.3 No Adjustment Under Certain Circumstances. No adjustment or action described in this Article VIII or in any other provision of the Plan shall be authorized to the extent that such adjustment or action would cause the Plan to fail to satisfy the requirements of Section 423 of the Code.

8.4 No Other Rights. Except as expressly provided in the Plan, no Participant shall have any rights by reason of any subdivision or consolidation of shares of shares of any class, the payment of any dividend, any increase or decrease in the number of shares of shares of any class or any dissolution, liquidation, merger, amalgamation or consolidation of the Company or any other corporation. Except as expressly provided in the Plan or pursuant to action of the Administrator under the Plan, no issuance by the Company of shares of shares of any class, or securities convertible into shares of shares of any class, shall affect, and no adjustment by reason thereof shall be made with respect to, the number of Shares subject to outstanding rights under the Plan or the Purchase Price with respect to any outstanding rights.

#### **ARTICLE IX. AMENDMENT, MODIFICATION AND TERMINATION**

9.1 Amendment, Modification and Termination. The Administrator may amend, suspend or terminate the Plan at any time and from time to time; provided, however, that approval of the Company's shareholders shall be required to amend the Plan to: (a) increase the aggregate number, or change the type, of shares that may be sold pursuant to rights under the Plan under Section 3.1 (other than an adjustment as provided by Article VIII); (b) change the corporations or classes of corporations whose employees may be granted rights under the Plan; or (c) change the Plan in any manner that would cause the Plan to no longer be an "employee stock purchase plan" within the meaning of Section 423(b) of the Code.

9.2 Certain Changes to Plan. Without shareholder consent and without regard to whether any Participant rights may be considered to have been adversely affected, to the extent permitted by Section 423 of the Code, the Administrator shall be entitled to change the Offering Periods, limit the frequency and/or number of changes in the amount withheld from Compensation during an Offering Period, establish the exchange ratio applicable to amounts withheld in a currency other than U.S. dollars, permit payroll withholding in excess of the amount designated by a Participant in order to adjust for delays or mistakes in the Company's processing of withholding elections, establish reasonable waiting and adjustment periods and/or accounting and crediting procedures to ensure that amounts applied toward the purchase of Common Shares for each Participant properly correspond with amounts withheld from the Participant's Compensation, and establish such other limitations or procedures as the Administrator determines in its sole discretion to be advisable that are consistent with the Plan.

9.3 Actions In the Event of Unfavorable Financial Accounting Consequences. In the event the Administrator determines that the ongoing operation of the Plan may result in unfavorable financial accounting consequences, the Administrator may, in its discretion and, to the extent necessary or desirable, modify or amend the Plan to reduce or eliminate such accounting consequence including, but not limited to:

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(a) altering the Purchase Price for any Offering Period including an Offering Period underway at the time of the change in Purchase Price;

(b) shortening any Offering Period so that the Offering Period ends on a new Purchase Date, including an Offering Period underway at the time of the Administrator action; and

(c) allocating Shares.

Such modifications or amendments shall not require shareholder approval or the consent of any Participant.

9.4 Payments Upon Termination of Plan. Upon termination of the Plan, the balance in each Participant's Plan account shall be refunded as soon as practicable after such termination, without any interest thereon.

#### **ARTICLE X. TERM OF PLAN**

The Plan shall be effective on the Effective Date. The effectiveness of the Plan shall be subject to approval of the Plan by the shareholders of the Company within twelve months following the date the Plan is first approved by the Board. No right may be granted under the Plan prior to such shareholder approval. No rights may be granted under the Plan during any period of suspension of the Plan or after termination of the Plan.

#### **ARTICLE XI. ADMINISTRATION**

11.1 Administrator. Unless otherwise determined by the Board, the Administrator of the Plan shall be the Compensation Committee of the Board (or another committee or a subcommittee of the Board to which the Board delegates administration of the Plan) (such committee, the “*Committee*”). The Board may at any time vest in the Board any authority or duties for administration of the Plan.

11.2 Action by the Administrator. Unless otherwise established by the Board or in any charter of the Administrator, a majority of the Administrator shall constitute a quorum. The acts of a majority of the members present at any meeting at which a quorum is present and, subject to Applicable Law and the Bye-laws of the Company, acts approved in writing by a majority of the Administrator in lieu of a meeting, shall be deemed the acts of the Administrator. Each member of the Administrator is entitled to, in good faith, rely or act upon any report or other information furnished to that member by any officer or other employee of the Company or any Designated Subsidiary, the Company’s independent certified public accountants, or any executive compensation consultant or other professional retained by the Company to assist in the administration of the Plan.

11.3 Authority of Administrator. The Administrator shall have the power, subject to, and within the limitations of, the express provisions of the Plan and Applicable Laws:

(a) To determine when and how rights to purchase Common Shares shall be granted and the provisions of each offering of such rights (which need not be identical).

(b) To designate from time to time which Subsidiaries of the Company shall be Designated Subsidiaries, which designation may be made without the approval of the shareholders of the Company.

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(c) To construe and interpret the Plan and rights granted under it, and to establish, amend and revoke rules and regulations for its administration. The Administrator, in the exercise of this power, may correct any defect, omission or inconsistency in the Plan, in a manner and to the extent it shall deem necessary or expedient to make the Plan fully effective.

(d) To amend, suspend or terminate the Plan as provided in Article IX.

(e) Generally, to exercise such powers and to perform such acts as the Administrator deems necessary or expedient to promote the best interests of the Company and its Subsidiaries and to carry out the intent that the Plan be treated as an “employee stock purchase plan” within the meaning of Section 423 of the Code.

11.4 Decisions Binding. The Administrator’s interpretation of the Plan, any rights granted pursuant to the Plan, any subscription agreement and all decisions and determinations by the Administrator with respect to the Plan are final, binding, and conclusive on all parties.

## ARTICLE XII. MISCELLANEOUS

12.1 Restriction upon Assignment. A right granted under the Plan shall not be transferable other than by will or the applicable laws of descent and distribution, and is exercisable during the Participant’s lifetime only by the Participant. Except as provided in Section 12.4 hereof, a right under the Plan may not be exercised to any extent except by the Participant. The Company shall not recognize and shall be under no duty to recognize any assignment or alienation of the Participant’s interest in the Plan, the Participant’s rights under the Plan or any rights thereunder.

12.2 Rights as a Shareholder. With respect to Shares subject to a right granted under the Plan, a Participant shall not be deemed to be a shareholder of the Company, and the Participant shall not have any of the rights or privileges of a shareholder, until such Shares have been issued to the Participant or his or her nominee following exercise of the Participant’s rights under the Plan. No adjustments shall be made for dividends (ordinary or extraordinary, whether in cash securities, or other property) or distribution or other rights for which the record date occurs prior to the date of such issuance, except as otherwise expressly provided herein or as determined by the Administrator.

12.3 Interest. No interest shall accrue on the payroll deductions or contributions of a Participant under the Plan.

12.4 Designation of Beneficiary.

(a) A Participant may, in the manner determined by the Administrator, file a written designation of a beneficiary who is to receive any Shares and/or cash, if any, from the Participant’s account under the Plan in the event of such Participant’s death subsequent to a Purchase Date on which the Participant’s rights are exercised but prior to delivery to such Participant of such Shares and cash. In addition, a Participant may file a written designation of a beneficiary who is to receive any cash from the Participant’s account under the Plan in the event of such Participant’s death prior to exercise of the Participant’s rights under the Plan. If the Participant is married and resides in a community property state, a designation of a person other than the Participant’s spouse as his or her beneficiary shall not be effective without the prior written consent of the Participant’s spouse.

(b) Such designation of beneficiary may be changed by the Participant at any time by written notice to the Company. In the event of the death of a Participant and in the absence of a

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beneficiary validly designated under the Plan who is living at the time of such Participant’s death, the Company shall deliver such Shares and/or cash to the executor or administrator of the estate of the Participant, or if no such executor or administrator has been appointed (to the knowledge of the Company), the Company, in its discretion, may deliver such Shares and/or cash to the spouse or to any one or more dependents or relatives of the Participant, or if no spouse, dependent or relative is known to the Company, then to such other person as the Company may designate.

12.5 Notices. All notices or other communications by a Participant to the Company under or in connection with the Plan shall be deemed to have been duly given when received in the form specified by the Company at the location, or by the person, designated by the Company for the receipt thereof.

12.6 Equal Rights and Privileges. Subject to Section 5.7, all Eligible Employees will have equal rights and privileges under this Plan so that this Plan qualifies as an “employee stock purchase plan” within the meaning of Section 423 of the Code. Subject to Section 5.7, any provision of this Plan that is inconsistent with Section 423 of the Code will, without further act or amendment by the Company, the Board or the Administrator, be reformed to comply with the equal rights and privileges requirement of Section 423 of the Code.

12.7 Use of Funds. All payroll deductions received or held by the Company under the Plan may be used by the Company for any corporate purpose, and the Company shall not be obligated to segregate such payroll deductions.

12.8 Reports. Statements of account shall be given to Participants at least annually, which statements shall set forth the amounts of payroll deductions, the Purchase Price, the number of Shares purchased and the remaining cash balance, if any.

12.9 No Employment Rights. Nothing in the Plan shall be construed to give any person (including any Eligible Employee or Participant) the right to remain in the employ of the Company or any Parent or Subsidiary or affect the right of the Company or any Parent or Subsidiary to terminate the employment of any person (including any Eligible Employee or Participant) at any time, with or without cause.

12.10 Notice of Disposition of Shares. Each Participant shall give prompt notice to the Company of any disposition or other transfer of any Shares purchased upon exercise of a right under the Plan if such disposition or transfer is made: (a) within two years from the Enrollment Date of the Offering Period in which the Shares were purchased or (b) within one year after the Purchase Date on which such Shares were purchased. Such notice shall specify the date of such disposition or other transfer and the amount realized, in cash, other property, assumption of indebtedness or other consideration, by the Participant in such disposition or other transfer.

12.11 Governing Law. The Plan and any agreements hereunder shall be administered, interpreted and enforced under the internal laws of Delaware without regard to conflicts of laws thereof or of any other jurisdiction.

12.12 Electronic Forms. To the extent permitted by Applicable Law and in the discretion of the Administrator, an Eligible Employee may submit any form or notice as set forth herein by means of an electronic form approved by the Administrator. Before the commencement of an Offering Period, the Administrator shall prescribe the time limits within which any such electronic form shall be submitted to the Administrator with respect to such Offering Period in order to be a valid election.

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**KINIKSA PHARMACEUTICALS, LTD.  
2018 EMPLOYEE SHARE PURCHASE PLAN**

**OFFERING DOCUMENT**

This document (this “*Offering Document*”) is adopted by the Compensation Committee of the Board of Directors of Kiniksa Pharmaceuticals, Ltd. (the “*Company*”), in its capacity as Administrator of the Kiniksa Pharmaceuticals, Ltd. 2018 Employee Share Purchase Plan (the “*Plan*”) and is made a part of the Plan. A copy of this Offering Document shall be attached to the Plan. Defined terms used in this Offering Document without definition have the meanings specified in the Plan.

This Offering Document shall apply to Offering Periods under the Plan until this Offering Document is terminated, amended or modified by the Administrator or a new Offering Document is adopted by the Administrator.

<b>Eligibility Requirements:</b>	Eligible Employees of the Company and the Designated Subsidiaries indicated below shall be eligible to participate, provided they meet the other eligibility requirements set forth in the Plan.
<b>Designated Subsidiaries:</b>	All Subsidiaries of the Company.
<b>Offering Periods to Commence:</b>	On each January 1 and July 1; <u>provided, however</u> , that the initial Offering Period under the Plan (the “ <i>Initial Offering Period</i> ”) shall commence on July 23, 2018 (such date, the “ <i>Initial Enrollment Date</i> ”) and end December 31, 2018 (the “ <i>Initial Offering End Date</i> ”).
<b>Length of Offering Periods:</b>	Six months; <u>provided, however</u> , that the Initial Offering Period will commence on the Initial Enrollment Date and end on the Initial Offering End Date.
<b>Purchase Dates:</b>	The Purchase Date with respect to an Offering Period shall occur on the final Trading Day of the Offering Period.
<b>Purchase Price:</b>	On each Purchase Date, the purchase price for a Share will be 85% of the Fair Market Value of a Share on the Enrollment Date or on the Purchase Date, whichever is lower; <u>provided, however</u> , that the Purchase Price may be adjusted by the Administrator pursuant to the Plan; <u>provided, further</u> , that the Purchase Price shall not be less than the par value of a Share.
<b>Contributions:</b>	A Participant may elect to have up to 10% of the Participant’s Compensation deducted on each payday on an after-tax basis for use in purchasing Common Shares pursuant to the Plan and subject to the limitations on purchasing Common Shares thereunder.
<b>Enrollment:</b>	Eligible Employees must enroll in an Offering Period by

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delivering to the Company or its designee at least seven days prior to the first day of the Offering Period (or not later than 5:00 pm Eastern Time on July 20, 2018 for the Initial Offering Period only) a completed subscription agreement in the form provided by the Company (a “*Subscription Agreement*”).

<b>Changes in Contribution Rates:</b>	Participants may not change their rate of contributions during an Offering Period for such Offering Period, but may change their rate of contributions for future Offering Periods by delivering to the Company or its designee at least seven days prior to the first day of the Offering Period a completed Subscription Agreement.
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<b>Withdrawals:</b>	A Participant may withdraw from an Offering Period not less than seven days prior to the final day of the Offering Period.
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If a Participant withdraws from an Offering Period, the Participant may elect to participate again in any subsequent Offering Period so long as the Participant is still eligible to participate in the Plan.

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**AMENDED AND RESTATED EMPLOYMENT AGREEMENT**

This Amended and Restated Employment Agreement (this “Agreement”) is made and entered into as of May 29, 2018 (the “Effective Date”), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the “Company”), and Sanj K. Patel (the “Executive”).

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

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

WHEREAS, the Company and the Executive are party to an Amended and Restated Employment Agreement dated as of December June 29, 2017 (the “Original Agreement”); and

WHEREAS, the Company and the Executive wish to amend and restate the Original Agreement in accordance with the terms set forth herein and the Executive shall continue to serve as its Chief Executive Officer on the terms and conditions set forth in this Agreement, and the Executive wishes to continue such employment.

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

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

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

(b) “Cause” shall mean:

(i) The Executive’s gross negligence or willful misconduct in performance of his duties to the Company, where such gross negligence or willful misconduct has resulted in material damage to the Company or any of its Affiliates or successors; or

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

(iii) The Executive’s commission of a felony or crime involving moral turpitude; or

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

(v) The Executive’s failure to comply with lawful directives of the Parent Board (as defined below), which has caused damage to the Company or any of its Affiliates or successors.

(c) “Change in Control” shall mean:

(i) a sale of all or substantially all of the Parent’s assets, or

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

(iii) A change in the composition of the Parent Board such that the individuals who, as of the Effective Date, constitute the Parent Board (such Parent Board shall be hereinafter referred to as the “Incumbent Board”) cease for any reason to constitute at least a majority of the Parent Board; provided, however, that, for purposes of this Agreement, any individual who becomes a member of the Parent Board subsequent to the Effective Date, whose election, or nomination for election by the Parent’s shareholders, was approved by a vote of at least a majority of those individuals who are members of the Incumbent Board (or deemed to be such pursuant to this proviso) shall be considered as though such individual were a member of the Incumbent Board; or

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

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

(B) solely because the level of ownership held by any institutional investor or any affiliate thereof or any other person, or persons acting as a group (the “Subject Person”), exceeds the designated percentage threshold of the outstanding voting capital shares as a result of a repurchase or other acquisition of voting capital shares by the Parent reducing the number of shares outstanding, provided that if a Change in

Control would occur (but for the operating of this sentence) as a result of the acquisition voting capital shares by the Parent, and after such share acquisition, the Subject Person becomes the owner of any additional voting capital shares that, assuming the repurchase or other acquisition had not occurred, increases the percentage of the then outstanding voting capital shares owned by such Subject Person over the designated percentage threshold, then a Change in Control shall be deemed to occur.

- (d) “Code” shall mean the Internal Revenue Code of 1986, as amended, and the rules and regulations promulgated thereunder.
- (e) “Employee Benefit Plan” shall have the meaning ascribed to such term in Section 3(3) of ERISA, as amended from time-to-time.
- (f) “Founder Invention and Non-Disclosure Agreement” shall mean the Founder Invention and Non-Disclosure Agreement, dated as of September 16, 2015.

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(g) “Founder Non-Competition and Non-Solicitation Agreement” shall mean the Founder Non-Competition and Non-Solicitation Agreement, dated as of September 16, 2015.

(h) “Good Reason” shall mean any of the following, occurring without the Executive’s written consent:

(i) the assignment to the Executive of duties materially inconsistent with the Executive’s title, position, status, reporting relationships, authority, duties or responsibilities as contemplated by Section 3, or any other action by the Company which results in a diminution in the Executive’s title, position, status, reporting relationships, authority, duties or responsibilities; or

(ii) a requirement that the Executive relocate his primary reporting location to a location more than fifty (50) miles from the location of the Company’s offices in Lexington, Massachusetts as of the Effective Date; or

(iii) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b) 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith which are remedied by the Company promptly after receipt of notice thereof given by the Executive; or

(iv) a material diminution in the budget over which Executive has responsibility; or

(v) a breach by the Company of this Agreement between the Company and Executive.

(i) “Parent” shall mean the Company’s parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

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

2. Acceptance and Term. Executive hereby accepts employment subject to the terms and conditions set forth in this Agreement. Executive’s employment shall continue until terminated pursuant to Section 5 hereof (the “Term”).

3. Position, Duties and Responsibilities.

(a) During the Term, the Executive shall serve the Company as its Chief Executive Officer and shall report to the Board of Directors of the Parent (the “Parent Board”). During the Term, the Executive shall be employed by the Company on a full-time basis and shall perform the duties and responsibilities of his position.

(b) The Executive is currently in the role of Chairman of the Parent Board and the Company, and shall continue to serve as a member of the Parent Board and the Company. The Parent shall propose to the shareholders of the Parent Board at each appropriate annual meeting of such shareholders during Executive’s employment the reelection of the Executive as a director of the Parent Board, provided that the Executive is otherwise eligible for such election.

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(c) In addition, and without further compensation, the Executive shall serve as a director and/or officer of one or more of the Company’s Affiliates if so elected or appointed from time-to-time.

(d) During the Term, the Executive shall devote his full business time and his best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of his duties and responsibilities hereunder. The Executive shall not engage in any other business activity or serve in any industry, trade, professional, governmental or academic position during the term of this Agreement, except such activities as shall not interfere with the performance of his duties to the Company. Notwithstanding the foregoing, the Executive shall be entitled to attend to personal and family affairs and investments, be involved in not-for-profit, charitable and professional activities and serve on up to two (2) for-profit boards, provided that the foregoing does not, individually or in the aggregate, materially interfere with Executive’s responsibilities under this Agreement.

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

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

(a) Base Salary. During the Term, the Company shall pay the Executive at the rate of not less than \$750,000 per annum, payable in accordance with the payroll practices of the Company for its executives and subject to increase from time-to-time by the Parent Board, in its sole discretion (such base salary, as from time-to-time increased, the "Base Salary"). The Base Salary shall not be reduced at any time (including after any such increase) without the Executive's prior written consent, and the term Base Salary as used in this Agreement shall refer to Executive's Base Salary as so increased.

(b) Bonus Compensation. During the Employment Period, the Executive shall be paid an annual cash bonus ("Annual Bonus") with a minimum target level of 60% of Annual Base Salary (the "Target Bonus"). The applicable corporate and individual performance targets shall be determined by the Compensation Committee of the Parent Board (the "Compensation Committee") after consultation with the Executive, within the first ninety (90) days of each calendar year. The actual Annual Bonus for each calendar year shall be determined in good faith by the Compensation Committee based upon actual corporate and individual performance against established objective for such year (and, if determined by the Compensation Committee, may exceed the Target Bonus) and shall be payable in accordance with the procedures specified by the Compensation Committee; provided that the Annual Bonus shall be paid to Executive no later than March 15<sup>th</sup> of the calendar year immediately following the calendar year in which it was earned.

(c) Equity Participation. Executive shall be eligible to receive equity awards annually under the Parent's 2018 Incentive Award Plan or any other equity plan (collectively with the Parent's 2015 Equity Incentive Plan, the "Equity Plan"), and the award agreements related to such plans, as determined in the discretion of the Parent Board, commencing upon the Effective Date. Notwithstanding the provisions of the Equity Plan or any agreement or award, Executive shall be immediately 100% fully vested in all equity awards granted to him under this Agreement or any other agreement that vest solely based on the passage of time in the event of a Change in Control in which the

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award is not being assumed or substituted as provided for in the applicable Equity Plan (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(d) Vacation. During the Term, the Executive shall be entitled to earn vacation at the rate of five (5) weeks per year, to be taken at such times and intervals as shall be determined by the Executive, subject to the reasonable business needs of the Company. The Executive shall be entitled to carryover up to two (2) weeks of vacation into the following calendar year. Vacation shall otherwise be governed by the policies of the Company, as in effect from time-to-time.

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

(f) Business Expenses. The Company shall promptly pay or reimburse the Executive for all reasonable business expenses incurred or paid by the Executive in the performance of his duties and responsibilities hereunder, subject to reasonable substantiation and documentation, as may be specified by the Company from time-to-time.

5. Termination of Employment and Severance Benefits. The Executive's employment hereunder shall terminate under the following circumstances:

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

(b) Disability.

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

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

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

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

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the nature of such Cause. In the cases of Section 1(b)(i), 1(b)(iv), or 1(b)(v) above, the Company may not effectuate the termination for Cause unless and until: (i) the Company provides Executive with written notice setting forth in reasonable detail the nature for such Cause which must be given to Executive no later than the thirtieth (30) day following the initial commission of the acts or omissions constituting Cause, and (ii) Executive fails to cure the alleged conduct or events supporting Cause within thirty (30) days following Executive's receipt of such notice.

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

(e) By the Executive for Good Reason. The Executive may terminate his employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating his employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

(f) By the Executive Other than for Good Reason. The Executive may terminate his employment hereunder at any time upon sixty (60) days' notice to the Company. In the event of termination of the Executive pursuant to this Section 5(f), the Parent Board or the Company may elect to waive the period of notice, or any portion thereof. In the event that the Parent Board or the Company so waives some or all of the period of notice, the Company shall pay the Executive his Base Salary for the period so waived.

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

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

(b) Severance. In the event the Executive's employment terminates pursuant to Section 5(a), 5(b), 5(d) or 5(e) of this Agreement, in addition to Final Compensation, the Company (i) shall accelerate the vesting of all unvested equity awards that vest solely based on the passage of time either then held by, or previously granted to, Executive (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards) by eighteen (18) months (for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement); and (ii) shall pay the Executive (A) a lump sum equal to the Base Salary plus the Target Bonus divided by twelve (12), then multiplied by the number of months set forth in the Severance Period (as defined below) (such payment, the "Severance Payment"), (B) the Post-Termination Bonus (as defined below) and (C) an additional one-time bonus of \$25,000 (the "One-Time Bonus"). Subject to Sections 6(e) and 7(a) of this Agreement (i) the Severance Payment and the One-Time Bonus shall be paid on the sixtieth (60th) day following the date of termination

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and (ii) the Post-Termination Bonus shall be paid at the time provided in the applicable Bonus Plan or form of annual award issued thereunder, but in no event later than March 15 of the year following the year in which the Separation from Service occurs; provided that if the termination occurs during the twelve (12) month period following a Change in Control, (i) the Post-Termination Bonus shall be paid by the sixtieth (60th) day following the date of termination and (ii) notwithstanding the provisions of the Equity Plan, the Executive shall be immediately 100% fully vested in all unvested equity awards that vest solely based on the passage of time of Parent or its Affiliates (including, without limitation, restricted stock, restricted stock units, stock options or other equity-based awards, whether granted to or held by Executive either before or after the date of this Agreement, and for the avoidance of doubt, with any equity awards that vest in whole or in part based on the attainment of performance-vesting conditions being governed by the terms of the applicable award agreement).

(c) Severance Period. For the purposes of this Agreement, the "Severance Period" shall be twenty-four (24) months.

(d) Post-Termination Bonus. For the purposes of this Agreement, the "Post-Termination Bonus" shall be a pro-rata share of the Target Bonus for the year in which the termination occurs.

(e) Release of Claims. Executive's right to receive the payments and benefits set forth in Section 6(b) is conditioned on the Executive's signing and returning to the Company a general release of claims in the form provided by the Company at the time the Executive's employment is terminated (the "Employee Release"). The Executive must sign and return the Employee Release, if at all, by the deadline specified therein, which deadline shall in no event be later than the sixtieth (60th) calendar day following the termination date. The Employee Release shall take effect on the expiration of any revocation period specified therein, which shall be no longer than seven (7) days from the date of the Executive's signature

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

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

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7. Timing of Payments and Section 409A.

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

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

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

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

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

## 8. Confidentiality; Cooperation

(a) Confidentiality and Other Covenants. As a condition of Executive's employment with the Company, the Executive has executed the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement. The parties hereto acknowledge and agree that this Agreement, the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement shall be considered separate contracts. In addition, Executive represents and warrants that he shall be able to and will perform the duties of this position without utilizing any material confidential and/or proprietary information that Executive may have obtained in connection with employment with any prior employer, and that he shall not (i) disclose any such information to the Company, or (ii) induce any Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement or otherwise.

(b) Litigation and Regulatory Cooperation. During and after Executive's employment, Executive shall reasonably cooperate with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Company employed Executive, provided, that the Executive will not have an obligation under this paragraph with respect to any claim in which the Executive has filed directly against the Company or related persons or entities. The Executive's reasonable cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after Executive's employment, Executive also shall reasonably

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cooperate with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while Executive was employed by the Company, provided Executive will not have any obligation under this paragraph with respect to any claim in which Executive has filed directly against the Company or related persons or entities. The Company shall reimburse Executive for any reasonable out-of-pocket expenses incurred in connection with Executive's performance of obligations pursuant to this Section 8(b).

## 9. Section 280G; Limitations on Payment

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

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

(c) Unless Executive and the Company agree on an alternative accounting firm or law firm, the accounting firm engaged by the Company for general tax compliance purposes as of the day prior to the effective date of the change of control transaction shall perform the foregoing calculations. If the accounting firm so engaged by the Company is serving as accountant or auditor for the individual, entity or group effecting the change of

control transaction, the Company shall appoint a nationally recognized accounting or law firm to make the determinations required by this Section 9. The Company shall bear all expenses with respect to the determinations by such accounting or law firm required to be made hereunder. The Company shall use commercially reasonable efforts to cause the accounting or law firm engaged to make the determinations hereunder to provide its calculations, together with detailed supporting documentation, to Executive and the Company within fifteen (15) calendar days after the date on which Executive's right to a 280G Payment becomes reasonably likely to occur (if requested at that time by Executive or the Company) or such other time as requested by Executive or the Company.

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

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

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

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

12. Assignment.

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

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

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

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

15. Notices. Any and all notices, requests, demands and other communications provided for by this Agreement shall be in writing and shall be effective when delivered in person, consigned to a

reputable national courier service or deposited in the United States mail, postage prepaid, registered or certified, and addressed to the Executive at his last known address on the books of the Company or, in the case of the Company, at its principal place of business, attention of the Compensation Committee of the Parent Board with a copy to the attention of the Chief Legal Officer, or to such other address as either party may specify by notice to the other actually received. Any notice so addressed shall be deemed to be given or received (a) if delivered by hand, on the date of such delivery, (b) if mailed by courier or by overnight mail, on the first business day following the date of such mailing, and (c) if mailed by registered or certified mail, on the third business day after the date of such mailing.

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement, and the Founder Non-Solicitation and Non-Competition Agreement, constitutes the entire understanding and agreement of the parties hereto regarding the employment of Executive. This Agreement supersedes all prior negotiations, discussions, correspondence, communications, understandings, and agreements between the parties (including any offer letter given to Executive) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Executive acknowledge that the Executive holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Executive under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

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

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

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

20. Governing Law. This is a Massachusetts contract and shall be construed and enforced under and be governed in all respects by the laws of the Commonwealth of Massachusetts, without regard to the conflict of laws principles thereof.

[Signature page follows immediately.]

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

**EXECUTIVE**

**KINIKSA PHARMACEUTICALS CORP.**

/s/ Sanj K. Patel  
Sanj K. Patel

By: /s/ Thomas W. Beetham  
Name: Thomas W. Beetham  
Title: Executive Vice President, Chief Legal Officer and Corporate Development

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**AMENDED AND RESTATED EMPLOYMENT AGREEMENT**

This Amended and Restated Employment Agreement (this “Agreement”) is made and entered into as of May 29, 2018 (the “Effective Date”), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the “Company”), and Stephen F. Mahoney (the “Employee”).

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

WHEREAS, the Employee is currently serving as the Company’s President and Chief Operating Officer and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the “Original Agreement”); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

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

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

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

(b) “Cause” shall mean:

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

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

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

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

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

*EVP (Founder)*

(c) “Change in Control” shall mean:

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

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

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

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

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

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

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

(f) “Founder Invention and Non-Disclosure Agreement” shall mean the Founder Invention and Non-Disclosure Agreement between the Employee and Company dated as of September 18, 2015.

(g) “Founder Non-Competition and Non-Solicitation Agreement” shall mean the Founder Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of September 18, 2015.

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(h) “Good Reason” shall mean any of the following, occurring without the Employee’s written consent:

(i) a demotion of the Employee to a position with responsibilities substantially less than the Employee’s prior position; provided that, a change in title, reporting relationships and/or responsibilities of the Employee could, but do not necessarily in and of themselves, individually or in the aggregate, constitute a demotion for purposes of this Section 1(h)(i); and in all instances, the determination of whether a demotion has occurred shall be made by the Company in good faith; or

(ii) a requirement that the Employee relocate Employee’s primary reporting location to a location more than fifty (50) miles from the location of the Company’s current offices in Lexington, Massachusetts as of the Effective Date; or

(iii) a reduction of more than five percent (5%) of Employee’s Base Salary, other than in connection with a reduction of similar magnitude to the base salaries of employees who are similarly situated to Employee; or

(iv) any failure by the Company to comply with any of the provisions of Section 4(a), 4(b), 4(c) or 4(d) hereof, other than insubstantial or inadvertent failures not in bad faith that are remedied by the Company promptly after receipt of notice thereof given by the Employee; or

(v) a breach by the Company of this Agreement.

(i) “Parent” shall mean the Company’s parent entity, Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company.

(j) “Parent Board” shall mean the board of directors of the Parent.

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

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

3. Position, Duties and Responsibilities.

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

(b) During the Term, the Employee shall devote Employee’s full business time and Employee’s best efforts, business judgment, skill and knowledge exclusively to the advancement of the business and interests of the Company and its Affiliates and to the discharge of Employee’s duties and responsibilities hereunder. During the Term, the Employee shall not engage in any other business

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activity or serve in any industry, trade, professional, governmental or academic position unless Employee first has obtained consent from the Chief Executive Officer of the Company.

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

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

(a) Base Salary. The Company shall pay the Employee an annual base salary of \$442,260, payable in accordance with the Company’s standard payroll practices and procedures and subject to change from time-to-time in the Company’s sole discretion (such base salary, as from time-to-time changed, the “Base Salary”).

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus (“Discretionary Annual Bonus”) with an initial target level of 40% of Employee’s Base Salary (the “Target Bonus”). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year,

if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15<sup>th</sup> of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

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

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

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

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

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

(b) Disability.

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

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

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

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

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

(e) By the Employee for Good Reason. The Employee may terminate Employee's employment hereunder for Good Reason (i) by providing notice to the Company specifying in reasonable detail the condition giving rise to the Good Reason no later than thirty (30) days following the occurrence of that condition; (ii) by providing the Company a period of thirty (30) days to remedy the condition and so specifying in the notice; and (iii) by terminating Employee's employment for Good Reason within thirty (30) days following the expiration of the period to remedy if the Company fails to remedy the situation.

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

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

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

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

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

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

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

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

## 7. **Timing of Payments and Section 409A.**

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

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

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

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

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

## 8. **Confidentiality; Cooperation**

(a) **Confidentiality and Other Covenants.** As a condition of Employee's employment with the Company, the Employee has executed the Founder Invention and Non-Disclosure Agreement and the Founder Invention and Non-Disclosure Agreement, both of which the Company and Employee acknowledge and agree shall be considered separate contracts. In addition, Employee represents and warrants that Employee shall be able to and will continue to perform the duties of Employee's position without utilizing any material confidential and/or proprietary information that Employee may have obtained in connection with employment with any prior employer, and that Employee shall not (i) disclose any such information to the Company, or (ii) induce any

Company employee to use any such information, in either case in violation of any confidentiality obligation, whether by agreement, by operation of law or otherwise.

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

9. Section 280G; Limitations on Payment

(a) If any payment or benefit Employee shall or may receive from the Company or otherwise (a "280G Payment") would (i) constitute a "parachute payment" within the meaning of Section 280G of the Code, and (ii) but for this sentence, be subject to the excise tax imposed by Section 4999 of the Code (the "Excise Tax"), then any such 280G Payment provided pursuant to this Agreement (a "Payment") shall be equal to the Reduced Amount. The "Reduced Amount" shall be either (x) the largest portion of the Payment that would result in no portion of the Payment (after reduction) being subject to the Excise Tax or (y) the largest portion, up to and including the total, of the Payment, whichever amount (i.e., the amount determined by clause (x) or by clause (y)), after taking into account all applicable federal, state and local employment taxes, income taxes, and the Excise Tax (all computed at the highest applicable marginal rate), results in Employee's receipt, on an after-tax basis, of the greater economic benefit notwithstanding that all or some portion of the Payment may be subject to the Excise Tax. If a reduction in a Payment is required pursuant to the preceding sentence and the Reduced Amount is determined pursuant to clause (x) of the preceding sentence, the

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reduction shall occur in the manner (the "Reduction Method") that results in the greatest economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the "Pro Rata Reduction Method").

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

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

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

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

10. Indemnification. The Company shall indemnify the Employee to the extent provided in its then current Certificate of Incorporation or By-Laws. The Employee agrees to promptly notify the Company of any actual or threatened claim arising out of or as a result of Employee's employment with the Company. The parties acknowledge that the Employee is also indemnified by the Parent to the extent set forth in the Indemnification Agreement between the Parent and Employee dated December 16, 2015.

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

12. Assignment.

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

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

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

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

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

16. Entire Agreement. This Agreement, together with the Founder Invention and Non-Disclosure Agreement and the Founder Non-Solicitation and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior

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negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that the Employee holds restricted stock of the Parent subject to a Restricted Stock Agreement and that options and other equity awards have been and, subject to the discretion and approval of the Parent Board, may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

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

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

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

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

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

EMPLOYEE

KINIKSA PHARMACEUTICALS CORP.

/s/ Stephen F. Mahoney

By: /s/ Sanj K. Patel



**AMENDED AND RESTATED EMPLOYMENT AGREEMENT**

This Amended and Restated Employment Agreement (this “Agreement”) is made and entered into as of May 29, 2018 (the “Effective Date”), by and between Kiniksa Pharmaceuticals Corp., a Delaware corporation (the “Company”), and John F. Paolini (the “Employee”).

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

WHEREAS, the Employee is currently serving as the Company’s Senior Vice President and Chief Medical Officer and possesses certain experience and expertise that qualify Employee to provide the direction and leadership required by the Company and its Affiliates;

WHEREAS, the Company and the Employee are party to an Employment Agreement dated as of November 1, 2016 (the “Original Agreement”); and

WHEREAS, the Company and the Employee wish to amend and restate the Original Agreement in accordance with the terms and conditions set forth in this Agreement.

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

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

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

(b) “Cause” shall mean:

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

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

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

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

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

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(c) “Change in Control” shall mean:

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

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

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

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

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

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

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

(f) “Confidential Information and Non-Competition Agreement” shall mean the Employee Proprietary Information, Inventions Assignment, Non-Competition and Non-Solicitation Agreement between the Employee and Company dated as of August 15, 2016.

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

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

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

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

3. Position, Duties and Responsibilities.

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

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

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

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

(a) Base Salary. The Company shall pay the Employee an annual base salary of \$420,000, payable in accordance with the Company’s standard payroll practices and procedures and subject to change from time-to-time in the Company’s sole discretion (such base salary, as from time-to-time changed, the “Base Salary”).

(b) Discretionary Bonus Compensation. During the Term, the Employee shall be eligible to receive an annual cash bonus (“Discretionary Annual Bonus”) with a target level of 30% of Employee’s Base Salary (the “Target Bonus”). The applicable performance goals shall be determined by the Company as soon as practicable at the beginning of each calendar year. The actual Discretionary Annual Bonus for each calendar year, if any, shall be determined in the sole and absolute discretion of the Company and shall be paid to Employee no later than March 15<sup>th</sup> of the calendar year immediately following the calendar year in which it was earned. For the avoidance of doubt, the Company reserves the right to not pay any Discretionary Annual Bonuses even if all performance goals are achieved or exceeded.

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

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

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

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

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

(b) Disability.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

7. Timing of Payments and Section 409A.

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

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

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

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

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

8. Confidentiality; Cooperation

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

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

9. Section 280G; Limitations on Payment

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

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

economic benefit for Employee. If more than one method of reduction shall result in the same economic benefit, the items so reduced shall be reduced pro rata (the “Pro Rata Reduction Method”).

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

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

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

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

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

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

12. Assignment.

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

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

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

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

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

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

16. Entire Agreement. This Agreement, together with the Confidential Information and Non-Competition Agreement, constitute the entire understanding and agreement of the Company and the Employee regarding the terms and conditions of Employee's employment with the Company. This Agreement supersedes all prior negotiations, discussions, correspondence, communications, understandings, and agreements between the Company and the Employee (including any offer letter given to Employee) relating to the subject matter of this Agreement, including (without limitation) the Original Agreement. Notwithstanding the foregoing, the Company and the Employee acknowledge that options and other equity awards have been and may be granted to Employee under and pursuant to the Parent's 2015 Equity Incentive Plan and any amendments thereto, as well as additional grants under the Parent's 2018 Incentive Award Plan or any additional equity plans of the Parent or its Affiliates, and the award agreements related to such plans (collectively, the "Awards"); and to the extent that the terms of this Agreement (including without limitation, Section 6(b)) accelerate the vesting of any such Awards, then the terms of this Agreement are intended to be in addition to the vesting provisions of such Awards and are not intended to diminish any vesting rights contained in such Awards.

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

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

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

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

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

**EMPLOYEE**

**KINIKSA PHARMACEUTICALS CORP.**

/s/ John F. Paolini

Name: John F. Paolini

By: /s/ Sanj K. Patel

Name: Sanj K. Patel

Title: Chief Executive Officer

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## FIRST AMENDMENT TO SUBLEASE AGREEMENT

**THIS FIRST AMENDMENT TO SUBLEASE AGREEMENT** (the “Amendment”) made as of June 26, 2018, by and between **SHIRE HUMAN GENETIC THERAPIES, INC.**, a Delaware corporation (the “Sublessor”) and **KINIKSA PHARMACEUTICALS CORP.**, a Delaware corporation (the “Sublessee”).

BACKGROUND

A. AMAG Pharmaceuticals, Inc., a Delaware corporation (“AMAG”), Sublessor’s predecessor in interest, and The Trustees of the 92 Hayden Avenue Trust, under Declaration of Trust dated August 18, 1983 (“Prime Lessor”), entered into a Lease dated May 22, 2008, as amended by a Consent to Assignment Agreement and Amendment to Lease among AMAG, Prime Lessor and Sublessor dated June 10, 2013 and a First Amendment to Lease dated November 24, 2015 (as amended, the “Prime Lease”), pursuant to which Prime Lessor as landlord leased to Sublessor as tenant the three (3) story building on the site known as 100 Hayden Avenue, Lexington, Massachusetts, containing approximately 55,924 square feet of rentable floor area as shown on Exhibit D of the Prime Lease (the “Leased Premises”) located on the Land as described in the Prime Lease; and

B. The Prime Lease is scheduled to expire on August 31, 2021; and

C. Sublessor and Sublessee entered into a Sublease Agreement dated as of March 13, 2018 (the “Sublease”), pursuant to which Sublessee subleased a portion of the Leased Premises consisting of approximately 25,067 rentable square feet, consisting of the entire third (3rd) floor (approximately 20,969 rentable square feet) and approximately 4,098 rentable square feet on the first floor in the Building as further described in the Sublease (together, the “Subleased Premises”).

D. Sublessor and Sublessee desire to amend the Sublease to add to the Subleased Premises approximately 2,177 rentable square feet, consisting of 2,157 located on the first floor of the Building and 20 rentable square feet located on the second floor of the Building, as delineated on **Exhibit A** attached hereto (the “Expansion Space”) which shall increase the rentable area of the Subleased Premises from 25,067 rentable square feet to 27,244 rentable square feet.

E. Capitalized terms not defined in this Amendment shall have the meaning provided by the Sublease.

**NOW, THEREFORE**, for and in consideration of the covenants and agreements set forth in this Amendment, and intending to be legally bound, Sublessor and Sublessee agree as follows:

1. **Demise; Term.**

1.1. Demise. Sublessor hereby subleases the Expansion Space to Sublessee, and Sublessee hereby subleases the Expansion Space from Sublessor, together with all fixtures installed in the Expansion Space by or for the benefit of Sublessor, and also together with all appurtenances and rights ancillary to the Expansion Space. The Expansion Space is subleased to Sublessee on the same terms as provided in the Sublease except as otherwise expressly provided herein. From and after the Expansion Space Commencement Date the Subleased Premises shall include the Expansion Space and Sublessee’s Share shall increase from 44.8% to 48.72%. and Sublessee shall be entitled to use ninety (90) parking spaces for Sublessee’s exclusive use.

1.2. Term. The term of the sublease of the Expansion Space to Sublessee shall begin (the “Expansion Premises Commencement Date”) on the date that this Amendment has been executed and delivered by Sublessor and Sublessee and that Sublessor and Sublessee have received the Prime Lessor’s Consent (as defined in Section 6.1 of this Amendment) and shall be coterminous with the Sublease Term.

1.3. Early Occupancy. Sublessee shall have the right to occupy the Expansion Premises from and after the Expansion Premises Commencement Date without being obligated to pay any Monthly Base Rent prior to Base Rent Commencement Date but Sublessee shall pay Additional Rent beginning on the date of early occupancy.

2. **Rent.**

Section 2.1 of the Sublease is deleted and replaced with the following:

2.1. Monthly Base Rent. Sublessee shall pay rent (“Monthly Base Rent”) as follows during the Sublease Term. Beginning on the August 1, 2018 (the “Base Rent Commencement Date”), Sublessee shall pay rent on the original 25,067 rentable square foot portion of the Subleased Premises in an amount equal to Sixty-Six Thousand Eight Hundred Forty-Five and 33/100 Dollars (\$66,845.33) per month. Beginning on October 1, 2018, Sublessee shall pay rent on the full 27,244 rentable square foot Subleased Premises in an amount equal to Seventy-Two Thousand Six Hundred Fifty and 66/100 Dollars (\$72,650.66) per month, and not later than the first (1<sup>st</sup>) day of each month thereafter.

3. **Expansion Premises Accepted “AS-IS”.**

Sublessor hereby represents and warrants to Sublessee that the Expansion Premises and all systems servicing the Expansion Premises are in good working order and condition, reasonable wear and tear excepted. Sublessor shall deliver the Expansion Premises broom clean, which Sublease Premises are accepted by Sublessee in their present condition, “**AS-IS**,” without any representation or warranty by Sublessor, subject to the state of title on the date of this Amendment, and also subject to all applicable legal requirements and any violation of legal requirements which may exist on the date of this Amendment. Sublessee has examined and approved the Expansion Premises and acknowledges that all improvements and fixtures included in the Expansion Premises are in good condition and working order. Sublessor shall

have no obligation to make any improvements to the Expansion Premises or provide Sublessee any allowance for so doing.

4. **Security Deposit.** \$220,000

5. **Tenant's Work and Available Allowance.**

Tenant's Work shall include the improvements described in **Exhibit B** hereto. The Available Allowance shall be increased from \$250,670.00 to \$272,440.00.

6. **Prime Lessor's Consent.**

6.1. Sublease Conditioned Upon Consent. This sublease of the Expansion Space is subject to, and conditioned upon, Sublessor's obtaining the written consent of Prime Lessor to this Amendment (the "Prime Lessor's Consent").

6.2. Delivery of Information. Sublessee shall promptly deliver to Sublessor any information reasonably required by Prime Lessor (in connection with the Prime Lessor's Consent) with respect to the nature and operation of Sublessee's business and/or the financial condition of Sublessee.

6.3. Agreements for Benefit of Prime Lessor. Sublessor and Sublessee hereby agree, for the benefit of Prime Lessor, that neither this Amendment nor the Prime Lessor's Consent shall:

6.3.1. create privity of contract between Prime Lessor and Sublessee;

6.3.2. be deemed to amend the Prime Lease in any way (unless Prime Lessor shall have expressly agreed in writing to such amendment); or

6.3.3. be construed as a waiver of Prime Lessor's right to consent to any assignment of the Prime Lease by Sublessor or any further subletting of the Premises.

6.4. Fee. Any fee charged by Prime Lessor in connection with the Prime Lessor's Consent shall be paid by Sublessor.

6.5. Effect of Failure to Obtain Prime Lessor's Consent. If the Prime Lessor's Consent is required under the Prime Lease and Prime Lessor fails to consent to this Amendment within thirty (30) days after the execution and delivery of this Amendment by the parties, either Sublessor or Sublessee may terminate this Amendment by giving written notice to the other at any time thereafter, but before Prime Lessor grants such consent. Upon such termination, (A) Sublessor will return any prepaid rent to Sublessee, (B) this Amendment will become null and void, and (C) neither party will have any liability or obligation to the other under this Amendment.

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7. **Brokers.**

Sublessor and Sublessee represent and warrant to each other that no broker or finder other than TWRBJ and CBRE (the "Brokers"), were instrumental in arranging or bringing about this transaction and that there are no claims or rights for commissions, finders' fees or other compensation (collectively, "compensation") by any person or entity other than the Brokers. Sublessor shall be solely responsible for all compensation payable to the Brokers, pursuant to a separate agreement between Sublessor and Brokers. If any broker or finder asserts a claim for compensation based upon any actual or alleged contact, dealings or communication with Sublessor or Sublessee, then the party through whom such broker or finder makes its claim shall indemnify and hold the other party (the "Indemnified Party") harmless from and against any and all claims, damages, judgments, suits, liabilities, losses, costs and expenses (including without limitation, reasonable attorneys' fees and court costs) suffered or incurred by or brought against the Indemnified Party in connection with such claim for compensation.

8. **Miscellaneous.**

8.1. Interpretation of Sublease. The headings and captions in this Amendment are inserted for convenience of reference only and in no way define, describe or limit the scope or intent of this Amendment or any of its provisions. Where the context so requires, the use of the singular shall include the plural and vice versa and the use of the masculine shall include the feminine and the neuter.

8.2. Governing Law; Jurisdiction and Venue. This Amendment and the Sublease shall be governed by and construed in accordance with the laws of the Commonwealth of Massachusetts.

8.3. No Recording. Neither this Amendment nor any memorandum or short form thereof may be recorded by Sublessee.

8.4. Survival. Any covenants set forth in this Amendment which, by their nature, would reasonably be expected to be performed after the expiration or earlier termination of this Amendment, shall survive the expiration or earlier termination of this Amendment.

8.5. Counterparts. This Amendment may be executed in two or more counterparts, each of which shall be deemed an original, but all of which taken together shall constitute one and the same instrument.

8.6. Transmission of Sublease by Facsimile or PDF. The transmission of a signed counterpart of this Amendment by facsimile or by portable document file ("PDF") shall have the same force and effect as delivery of an original signed counterpart of this Amendment, and shall constitute valid and effective delivery for all purposes. If either party delivers a signed counterpart of this Amendment by transmission of a facsimile or PDF, it shall also send promptly thereafter by overnight courier or personal delivery a signed original counterpart of this Amendment to the other party, but failure to do so shall not render this Amendment void or voidable by either party.

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8.7. Binding Effect; Assignment. Subject to Article 7 of the Sublease, this Amendment shall be binding upon, and inure to the benefit of, the parties to this Amendment and their respective successors and assigns.

8.8. Entire Agreement; Requirement for Writing.

8.8.1. This Amendment and the Exhibits attached to this Amendment contain the final and entire agreement of Sublessor and Sublessee and are intended to be an integration of all prior negotiations and understandings. Neither Sublessor nor Sublessee shall be bound by any covenants, agreements, statements, representations or warranties, oral or written, not contained in this Amendment.

8.8.2. No change or modification to this Amendment shall be valid unless the same is in writing and signed by the parties to this Amendment.

8.8.3. No waiver of any of the provisions of this Amendment shall be valid unless the same is in writing and is signed by the party against which it is sought to be enforced.

8.9. Severability. If any provision of this Amendment, or the application thereof to any person, place or circumstance, shall be held by a court of competent jurisdiction to be invalid, unenforceable or void, the remainder of this Amendment and such provisions as applied to other persons, places and circumstances shall remain in full force and effect.

8.10. Time of Essence. Time is of the essence of each and every provision of this Amendment of which time is an element.

8.11. Drafts not an Offer to Enter into a Legally Binding Contract. The submission of a draft of this Amendment by one party to another is not intended by either party to be an offer to enter into a legally binding contract. The parties shall be legally bound pursuant to the terms of this Amendment only if and when Sublessor and Sublessee have fully executed and delivered to each other a counterpart of this Amendment.

8.12. Full Force and Effect. As modified by this Amendment the Sublease is in full force and effect.

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**IN WITNESS WHEREOF**, Sublessor and Sublessee have duly executed this Amendment as of the day and year first above written.

SUBLESSOR:

**SHIRE HUMAN GENETIC THERAPIES, INC.**

By: /s/ Michael Connor  
Name: Michael Connor  
Title: Head of Global Real Estate

SUBLESSEE:

**KINIKSA PHARMACEUTICALS CORP.**

By: /s/ Thomas Beetham  
Name: Thomas Beetham  
Title: EVP

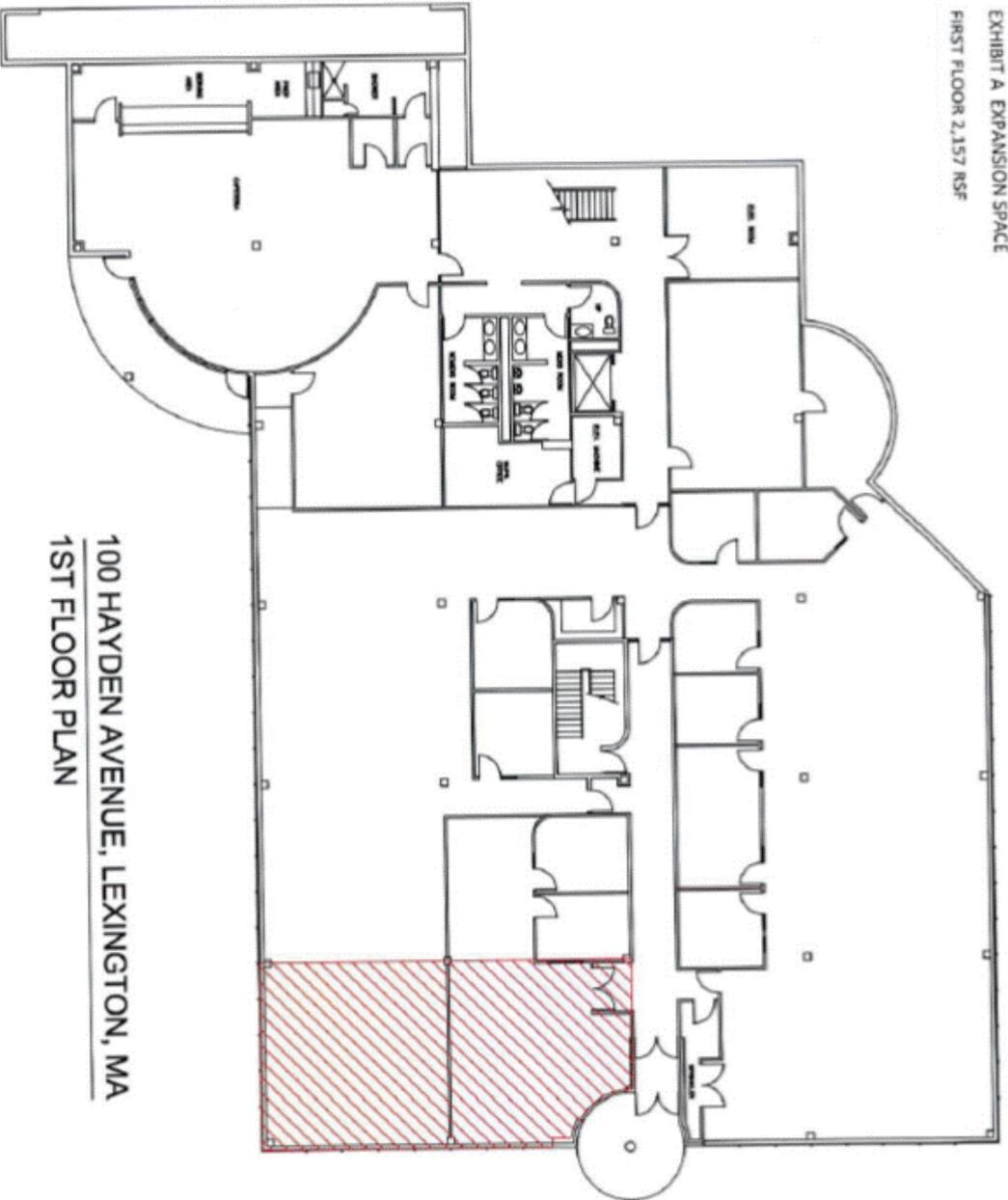
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**EXHIBIT A**

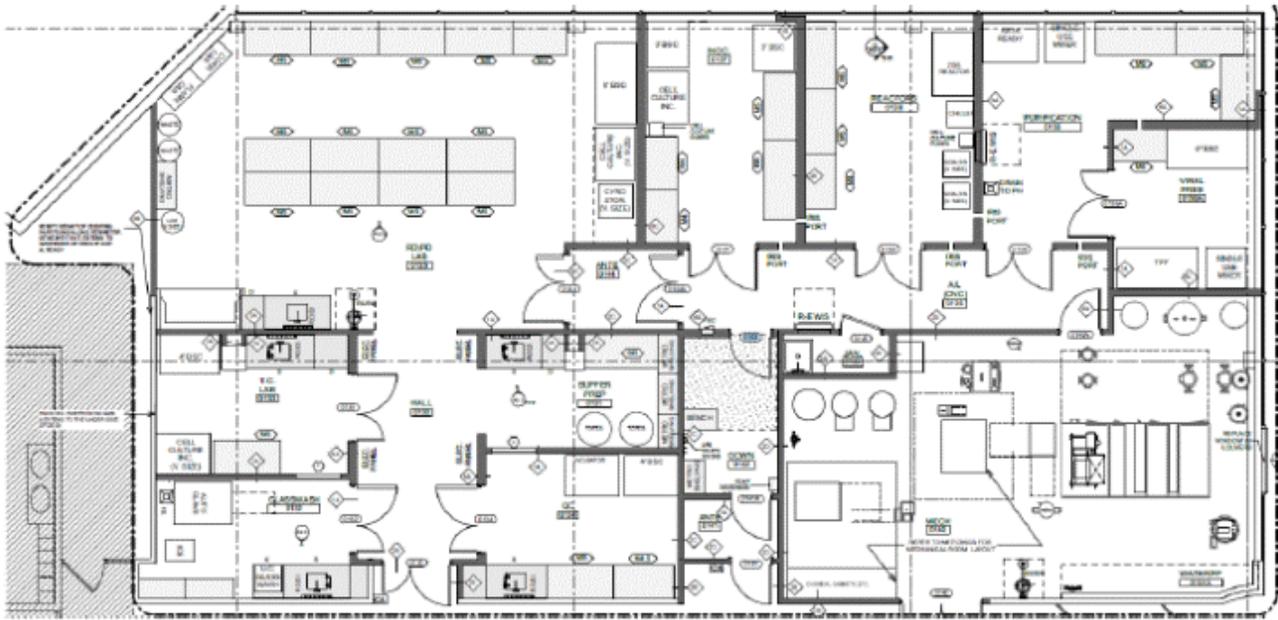
**EXPANSION SPACE**

EXHIBIT A EXPANSION SPACE  
FIRST FLOOR 2,157 RSF



100 HAYDEN AVENUE, LEXINGTON, MA  
1ST FLOOR PLAN





To support manufacturing and development activities, 962 sq/ft will be modified to allow for cold chain storage, equipment storage, and consumables. The common break room adjacent to the proposed storage room will not part of the construction activities.

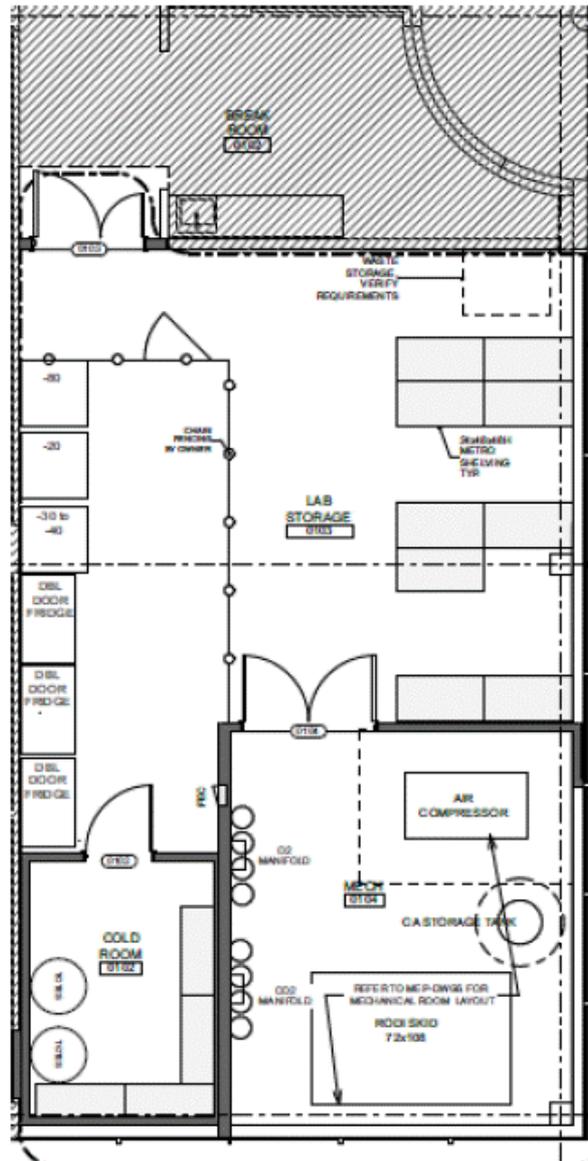


EXHIBIT C

## BILL OF SALE

**THIS SPECIAL WARRANTY BILL OF SALE (“Bill of Sale”)** is effective as of June 28, 2018, by **SHIRE HUMAN GENETIC THERAPIES, INC.**, a Delaware limited liability company (“**Seller**”) in favor **KINIKSA PHARMACEUTICALS CORP.**, a corporation (“**Buyer**”).

### BACKGROUND

**WHEREAS**, Seller has agreed to sell to Buyer and Buyer has agreed to purchase from Seller, all of Seller’s right, title and interest in and to all furniture, equipment and other personal property owned by Seller (the “**FF&E**”) located at 100 Hayden Avenue, Lexington, Massachusetts, and listed in Exhibit A attached hereto and made a part hereof (“**Subleased Premises**”).

**NOW, THEREFORE**, for and in consideration of \$1.00 and other good and valuable consideration, the receipt of which shall be a condition to the effectiveness of the following grant and conveyance, Seller does hereby grant, convey, transfer, bargain, sell, deliver and set over, all of Seller’s right, title and interest in and to the FF&E owned by Seller located at the Subleased Premises to Buyer, its successors and assigns.

Seller hereby warrants that Seller has the authority to transfer its interest in the FF&E and that Seller has good and marketable title to the FF&E, free and clear of all liens, claims, encumbrances and rights of other and that it will warrant and defend such title forever against all claims and demands whatsoever resulting from claims occurring prior to the effective date hereof.

Seller makes no representations, express or implied, as to the condition, usefulness or state of function or repair of the FF&E including without limitation, warranties of fitness or merchantability, it being expressly understood that the FF&E is being sold to Buyer “As is, Where is”, with all faults.

**IN WITNESS WHEREOF**, Seller has executed this Bill of Sale on the day and year first written above.

**SHIRE HUMAN GENETIC THERAPIES, INC.**

By: /s/ Michael Connor  
Name: Michael Connor  
Title: Head of Global Real Estate

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### EXHIBIT A TO BILL OF SALE

#### FF&E Inventory

In addition to the foregoing, the FF&E shall include all furniture, fixtures and equipment physically located in the Expansion Premises as of April , 2018.

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### SECOND AMENDMENT TO SUBLEASE AGREEMENT

**THIS SECOND AMENDMENT TO SUBLEASE AGREEMENT** (the “**Amendment**”) made as of July 17, 2018, by and between **SHIRE HUMAN GENETIC THERAPIES, INC.**, a Delaware corporation (the “**Sublessor**”) and **KINIKSA PHARMACEUTICALS CORP.**, a Delaware corporation (the “**Sublessee**”).

#### BACKGROUND

A. AMAG Pharmaceuticals, Inc., a Delaware corporation (“**AMAG**”), Sublessor’s predecessor in interest, and The Trustees of the 92 Hayden Avenue Trust, under Declaration of Trust dated August 18, 1983 (“**Prime Lessor**”), entered into a Lease dated May 22, 2008, as amended by a Consent to Assignment Agreement and Amendment to Lease among AMAG, Prime Lessor and Sublessor dated June 10, 2013 and a First Amendment to Lease dated November 24, 2015 (as amended, the “**Prime Lease**”), pursuant to which Prime Lessor as landlord leased to Sublessor as tenant the three (3) story building on the site known as 100 Hayden Avenue, Lexington, Massachusetts, containing approximately 55,924 square feet of rentable floor area as shown on Exhibit D of the Prime Lease (the “**Leased Premises**”) located on the Land as described in the Prime Lease; and

B. The Prime Lease is scheduled to expire on August 31, 2021; and

C. Sublessor and Sublessee entered into a Sublease Agreement dated as of March 13, 2018 (the “**Original Sublease**”), as amended by that certain First Amendment to Sublease dated as of June 26, 2018 (the “**First Amendment**” and collectively with the Original Sublease, the “**Sublease**”), pursuant to which Sublessee subleased a portion of the Leased Premises consisting of approximately 27,244 rentable square feet in the Building as further described in the Sublease (together, the “**Subleased Premises**”).

D. Sublessor and Sublessee desire to amend the Sublease to correct a scrivener’s error in the calculation of the electricity charges in Section 4.1.2 of the Original Sublease.

E. Capitalized terms not defined in this Amendment shall have the meaning provided by the Sublease.

**NOW, THEREFORE**, for and in consideration of the covenants and agreements set forth in this Amendment, and intending to be legally bound, Sublessor and Sublessee agree as follows:

1. **Electricity, Janitorial.**

Section 4.1.2 of the Original Sublease is deleted and replaced with the following:

“Electricity, Janitorial. Sublessor shall pay the costs of electrical service provided to the Sublease Premises determined as follows: (i) Sublessee at its costs shall cause the first floor Subleased Premises to be sub-metered and shall pay the costs of monthly meter readings and electrical costs for service to the first floor

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Subleased Premises; (ii) Sublessee shall pay the following amount each year for electrical service for the third floor Subleased Premises: \$3.10/square foot in equal monthly installments of \$5,416.99. Sublessee shall contract directly for, and pay the costs of, janitorial services for the laboratory portions of the Subleased Premises, and janitorial services for all other portions of the Subleased Premises shall be provided by Prime Lessor. Sublessee shall pay Sublessee’s Share of the costs of electrical service and common area janitorial services provided in the Building by Prime Lessor or Sublessor.”

2. **Miscellaneous.**

2.1. Interpretation of Sublease. The headings and captions in this Amendment are inserted for convenience of reference only and in no way define, describe or limit the scope or intent of this Amendment or any of its provisions. Where the context so requires, the use of the singular shall include the plural and vice versa and the use of the masculine shall include the feminine and the neuter.

2.2. Governing Law; Jurisdiction and Venue. This Amendment and the Sublease shall be governed by and construed in accordance with the laws of the Commonwealth of Massachusetts.

2.3. No Recording. Neither this Amendment nor any memorandum or short form thereof may be recorded by Sublessee.

2.4. Survival. Any covenants set forth in this Amendment which, by their nature, would reasonably be expected to be performed after the expiration or earlier termination of this Amendment, shall survive the expiration or earlier termination of this Amendment.

2.5. Counterparts. This Amendment may be executed in two or more counterparts, each of which shall be deemed an original, but all of which taken together shall constitute one and the same instrument.

2.6. Transmission of Sublease by Facsimile or PDF. The transmission of a signed counterpart of this Amendment by facsimile or by portable document file (“PDF”) shall have the same force and effect as delivery of an original signed counterpart of this Amendment, and shall constitute valid and effective delivery for all purposes. If either party delivers a signed counterpart of this Amendment by transmission of a facsimile or PDF, it shall also send promptly thereafter by overnight courier or personal delivery a signed original counterpart of this Amendment to the other party, but failure to do so shall not render this Amendment void or voidable by either party.

2.7. Binding Effect; Assignment. Subject to Article 7 of the Sublease, this Amendment shall be binding upon, and inure to the benefit of, the parties to this Amendment and their respective successors and assigns.

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2.8. Entire Agreement; Requirement for Writing.

2.8.1. This Amendment contains the final and entire agreement of Sublessor and Sublessee and are intended to be an integration of all prior negotiations and understandings. Neither Sublessor nor Sublessee shall be bound by any covenants, agreements, statements, representations or warranties, oral or written, not contained in this Amendment.

2.8.2. No change or modification to this Amendment shall be valid unless the same is in writing and signed by the parties to this Amendment.

2.8.3. No waiver of any of the provisions of this Amendment shall be valid unless the same is in writing and is signed by the party against which it is sought to be enforced.

2.9. Severability. If any provision of this Amendment, or the application thereof to any person, place or circumstance, shall be held by a court of competent jurisdiction to be invalid, unenforceable or void, the remainder of this Amendment and such provisions as applied to other persons, places and circumstances shall remain in full force and effect.

2.10. Time of Essence. Time is of the essence of each and every provision of this Amendment of which time is an element.

2.11. Drafts not an Offer to Enter into a Legally Binding Contract. The submission of a draft of this Amendment by one party to another is not intended by either party to be an offer to enter into a legally binding contract. The parties shall be legally bound pursuant to the terms of this Amendment only if and when Sublessor and Sublessee have fully executed and delivered to each other a counterpart of this Amendment.

2.12. Full Force and Effect. As modified by this Amendment the Sublease is in full force and effect.

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above written.

2.12.1. **IN WITNESS WHEREOF**, Sublessor and Sublessee have duly executed this Amendment as of the day and year first

SUBLESSOR:

**SHIRE HUMAN GENETIC THERAPIES, INC.**

By: /s/ Michael Connor

Name: Michael Connor

Title: Head of Global Real Estate

SUBLESSEE:

**KINIKSA PHARMACEUTICALS CORP.**

By: /s/ Thomas Beetham

Name: Thomas Beetham

Title: EVP

## CERTIFICATIONS

I, Sanj K. Patel, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Kiniksa Pharmaceuticals, Ltd.;
2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;
3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;
4. The registrant's other certifying officer(s) and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) for the registrant and have:
  - (a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;
  - (b) [OMITTED]
  - (c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and
  - (d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and
5. The registrant's other certifying officer(s) and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):
  - (a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and
  - (b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

August 6, 2018

/s/ Sanj K. Patel

Sanj K. Patel

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

## CERTIFICATIONS

I, Chris Heberlig, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Kiniksa Pharmaceuticals, Ltd.;
2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;
3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;
4. The registrant's other certifying officer(s) and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) for the registrant and have:
  - (a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;
  - (b) [OMITTED]
  - (c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and
  - (d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and
5. The registrant's other certifying officer(s) and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):
  - (a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and
  - (b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

August 6, 2018

/s/ Chris Heberlig  
Chris Heberlig  
Chief Financial Officer  
(Principal Financial Officer)

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

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

- (1) The Quarterly Report on Form 10-Q of the Company for the period ended June 30, 2018 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and
- (2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

August 6, 2018

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

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

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

- (1) The Quarterly Report on Form 10-Q of the Company for the period ended June 30, 2018 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and
- (2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

August 6, 2018

/s/ Chris Heberlig  
\_\_\_\_\_  
Chris Heberlig  
Chief Financial Officer  
(Principal Financial Officer)