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UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

FORM 10-Q

(Mark One)

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

For the quarterly period ended September 30, 2022
OR

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

For the transition period from to
Commission file number: 001-38492

Kiniksa Pharmaceuticals, Ltd.

(Exact Name of Registrant as Specified in Its Charter)

Bermuda	**98-1327726**
(State or Other Jurisdiction of Incorporation or Organization)	(I.R.S. Employer Identification No.)

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

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

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

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

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

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

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

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

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

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

As of October 31, 2022, there were 69,531,762 common shares outstanding in aggregate, comprised of:
34,531,084 Class A common shares, par value $0.000273235 per share
1,813,457 Class B common shares, par value $0.000273235 per share
17,129,603 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 NINE MONTHS ENDED SEPTEMBER 30, 2022

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SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This Quarterly Report on Form 10-Q, or Quarterly Report, contains forward-looking statements. All statements other than statements of historical facts contained in this Quarterly Report including statements regarding our products' commercial sales, future results of anticipated products, future results of operations and financial position, expected timeline for our cash, cash equivalents and short-term investments, business strategy, product development, prospective products and product candidates, their expected properties, performance, market opportunity and competition, supply of drug products at acceptable cost and quality, collaborators, license and other strategic arrangements, the expected timeline for achievement of our clinical milestones, the timing of, and potential results from, clinical and other trials, potential marketing authorization from the FDA or regulatory authorities in other jurisdictions, potential and ongoing coverage and reimbursement for our products and product candidates, if approved, commercial strategy and pre-commercial activities, research and development costs, timing of regulatory filings and feedback, timing and likelihood of success and plans and objectives of management for future operations and funding requirements, are forward-looking statements.

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

In some cases, you can identify forward-looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "goal," "design," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. The forward-looking statements in this Quarterly Report are only predictions. We have based these forward-looking statements largely on our current expectations and projections about future events and financial trends that we believe may affect our business, financial condition and results of operations. These forward-looking statements speak only as of the date of this Quarterly Report and are subject to a number of risks, uncertainties and assumptions described under the sections in this Quarterly Report entitled "Risk Factors" and "Management's Discussion and Analysis of Financial Condition and Results of Operations" and elsewhere in this Quarterly Report. These forward-looking statements are subject to numerous risks and uncertainties, including, without limitation, the following:

- our continued ability to commercialize ARCALYST (rilonacept) and to develop and commercialize our current and future product candidates, if approved;
- our status as an early-commercial stage biopharmaceutical company and our expectation to incur losses in the future;
- our future capital needs and our need to raise additional funds;
- our ability to manufacture sufficient commercial stock of our products to meet patient and partner demand;
- the market acceptance of our products and product candidates;
- competitive and potentially competitive products and technologies;
- prescriber awareness and adoption of our products and product candidates, if approved;
- the size of the market for our products and product candidates, if approved;
- our ability to meet the quality expectations of prescribers or patients;
- the decision of third-party payors not to cover or maintain coverage of or to establish burdensome requirements prior to covering ARCALYST or any of our current or future product candidates, if approved,

or to require extensive or independently performed clinical trials prior to covering or maintaining coverage of our product candidates, if approved;

- the lengthy and expensive clinical development process with its uncertain outcomes and potential for clinical failure or delay, including due to the COVID-19 pandemic and global political turmoil, including the ongoing war in Ukraine;
- the decision by any applicable regulatory authority whether to clear our current or future 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 products and product candidates;
- our ability to improve our product candidates;
- our ability to identify, in-license, acquire, discover or develop additional product candidates;
- our ability to undertake and execute on business combinations, collaborations or other strategic transactions;
- our ability to have our products and product candidates manufactured in accordance with regulatory requirements and at acceptable cost and quality specifications;
- 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;
- federal, state and foreign regulatory requirements applicable to our products and product candidates;
- our ability to obtain, maintain, protect and enforce our intellectual property rights related to our products and product candidates;
- the impact of the coronavirus disease 2019, or COVID-19, pandemic on our business, including our commercial operations and clinical trials;
- ownership concentration of our executive officers, directors, certain members of senior management and affiliated shareholders may prevent our shareholders from influencing significant corporate decisions; and
- our ability to attract and retain skilled personnel.

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

You should read this Quarterly Report and the documents that we reference in this Quarterly Report completely and with the understanding that our actual future results may be materially different from what we expect. We qualify all of our forward-looking statements by these cautionary statements.

SUMMARY RISK FACTORS

Our business is subject to numerous risks and uncertainties, including those described in Part II, Item 1A. “Risk Factors” in this Quarterly Report. You should carefully consider these risks and uncertainties when investing in our Class A common shares. The principal risks and uncertainties affecting our business include the following:

- we have only recently begun generating product revenue, have incurred significant operating losses in the past, expect to incur significant operating losses for the foreseeable future and may never achieve or maintain sustained corporate profitability;
- we will require significant additional funding to develop our portfolio, commercialize our products and product candidates, if approved, and to identify, discover, develop or acquire additional product candidates, and if we are unable to secure financing on acceptable terms when needed, or at all, we could be forced to delay, reduce or cease one or more of our product development plans, research and development programs or other operations or commercialization efforts;
- we depend heavily on the commercial success of ARCALYST, and have limited experience commercializing a therapeutic, supporting sales, marketing, and distribution activities and maintaining applicable infrastructure for these activities either directly and/or through agreements with third parties; as a result, we may not be able to sustain the commercialization of ARCALYST, or successfully commercialize any future approved product candidates;
- we depend heavily on the success of one or more of our product candidates, which are in various stages of product development; such success is dependent upon us advancing our product candidates in clinical development, obtaining regulatory approval and ultimately commercializing one or more of our product candidates on a timely basis, if at all;
- ARCALYST in recurrent pericarditis, as well as our current or future product candidates, if approved, may not gain sustained market acceptance by prescribers, patients, or third-party payors, in which case our ability to generate product revenues will be impaired;
- successful commercialization of our products and product candidates, if approved, will depend in part on the extent to which third-party payors provide funding, establish favorable coverage and pricing policies, respond to price increases and set adequate reimbursement levels for our products and product candidates, if approved, and failure to obtain or maintain coverage and adequate reimbursement for our products and product candidates, if approved, could limit our ability to market those products and decrease our ability to generate revenue;
- the incidence and prevalence for target patient populations of our products and product candidates have not been established with precision, and if the market opportunities for our products and product candidates, if approved, 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;
- clinical development of our product candidates is a lengthy and expensive process with uncertain timelines, costs and outcomes;
- we may encounter substantial delays in our current or planned preclinical and/or clinical trials, including as a result of delays in obtaining regulatory approvals for indications, site activation, enrollment, and conduct of the trials, which could delay or prevent our product development activities;
- we rely on third parties, including contract research organizations, or CROs, to activate our sites, conduct or otherwise support our research activities, preclinical studies and clinical trials for our product candidates, and these third parties may not perform satisfactorily, which could delay, prevent or impair our product development activities;

- we rely on third parties, including independent contract manufacturing organizations, or CMOs, to manufacture our product candidates for preclinical and clinical development, to manufacture our commercial supply of ARCALYST, and supply of drug substance and drug product for our products and product candidates; and if these third parties do not perform satisfactorily, including by producing insufficient commercial supply of ARCALYST to meet patient demand, or are impacted by delays or supply shortages, our product development activities, regulatory approval, and commercialization efforts may be delayed, prevented or impaired;

- the ongoing COVID-19 pandemic, and related measures taken in response, including measures imposed or re-imposed in light of new variants of the virus, may have an adverse impact on our business and operations as well as those of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including regulatory authorities;

- all of our products and product candidates have been licensed or acquired from other parties; if those parties did not adequately protect and we are unable to adequately protect such products and product candidates, or to secure and maintain freedom to operate, others could preclude us from commercializing our products and product candidates, if approved, or compete against us more directly;

- we face significant competition from other biotechnology and pharmaceutical companies, which may result in others discovering, developing or commercializing drugs before or more successfully than us;

- we may not successfully execute our growth strategy to identify, discover, develop, license or acquire additional product candidates or technologies, and our strategy may not deliver anticipated results or we may refine or otherwise alter our growth strategy;

- we may seek to acquire businesses or undertake business combinations, collaborations or other strategic transactions which may not be successful or on favorable terms, if at all, and we may not realize the intended benefits of such transactions; and

- concentration of ownership of the voting power of our common shares may prevent new investors from influencing significant corporate decisions and may have an adverse effect on the price of our Class A common shares.

INDUSTRY AND OTHER DATA

Unless otherwise indicated, certain industry data and market data included in this Quarterly Report were obtained from independent third-party surveys, market research, publicly available information, reports of governmental agencies and industry publications and surveys. All of the market data used in this Quarterly Report involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. We believe that the information from these industry publications and surveys included in this Quarterly Report is reliable.

ARCALYST is a registered trademark of Regeneron Pharmaceuticals, Inc. (“Regeneron”). Solely for convenience, trademarks, service marks, and trade names referred to in this Quarterly Report may be listed without identifying symbols.

Part I — Financial Information

Item 1. Financial Statements (unaudited)

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

	September 30, 2022	December 31, 2021
Assets		
Current assets:		
Cash and cash equivalents	$ 175,761	$ 122,470
Short-term investments	24,963	59,731
Accounts receivable, net	11,159	3,985
Inventory	14,621	3,675
Prepaid expenses and other current assets	15,311	6,585
Total current assets	241,815	196,446
Property and equipment, net	1,995	2,834
Operating lease right-of-use assets	6,173	5,550
Other long-term assets	5,627	8,720
Intangible asset, net	18,500	19,250
Deferred tax assets	185,843	—
Total assets	$ 459,953	$ 232,800
Liabilities and Shareholders' Equity		
Current liabilities:		
Accounts payable	$ 1,469	$ 1,868
Accrued expenses	30,868	38,031
Deferred revenue	12,092	—
Operating lease liabilities	3,274	3,381
Other current liabilities	8,789	1,544
Total current liabilities	56,492	44,824
Non-current liabilities:		
Non-current deferred revenue	14,198	—
Non-current operating lease liabilities	3,435	2,669
Other long-term liabilities	1,820	270
Total liabilities	75,945	47,763
Commitments and contingencies (Note 14)		
Shareholders' equity:		
Class A common shares, par value of $0.000273235 per share; 34,523,660 shares and 34,059,725 shares issued and outstanding as of September 30, 2022 and December 31, 2021, respectively	9	8
Class B common shares, par value of $0.000273235 per share; 1,813,457 shares issued and outstanding as of September 30, 2022 and December 31, 2021	1	1
Class A1 common shares, $0.000273235 par value; 17,129,603 shares issued and outstanding as of September 30, 2022 and December 31, 2021	5	5
Class B1 common shares, $0.000273235 par value; 16,057,618 shares issued and outstanding as of September 30, 2022 and December 31, 2021	4	4
Additional paid-in capital	880,554	860,482
Accumulated other comprehensive loss	(70)	(66)
Accumulated deficit	(496,495)	(675,397)
Total shareholders' equity	384,008	185,037
Total liabilities and shareholders' equity	$ 459,953	$ 232,800

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

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

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2022	2021	2022	2021
Revenue:				
Product revenue, net	$ 33,424	$ 12,095	$ 82,585	$ 19,799
License and collaboration revenue	65,711	—	75,711	—
Total revenue	99,135	12,095	158,296	19,799
Costs and operating expenses:				
Cost of goods sold	6,937	2,767	16,185	5,233
Collaboration expenses	4,623	—	16,549	—
Research and development	16,485	19,236	51,100	71,864
Selling, general and administrative	24,677	20,759	70,736	63,207
Total operating expenses	52,722	42,762	154,570	140,304
Income (loss) from operations	46,413	(30,667)	3,726	(120,505)
Interest income	322	5	459	20
Income (loss) before income taxes	46,735	(30,662)	4,185	(120,485)
Benefit (provision) for income taxes	177,358	118	174,717	(1,106)
Net income (loss)	$ 224,093	$ (30,544)	$ 178,902	$ (121,591)
Net income (loss) per share attributable to common shareholders—basic	$ 3.23	$ (0.44)	$ 2.58	$ (1.78)
Net income (loss) per share attributable to common shareholders—diluted	$ 3.18	$ (0.44)	$ 2.55	$ (1.78)
Weighted average common shares outstanding—basic	69,445,071	68,662,673	69,305,755	68,444,061
Weighted average common shares outstanding—diluted	70,552,018	68,662,673	70,286,444	68,444,061
Comprehensive income (loss):				
Net income (loss)	$ 224,093	$ (30,544)	$ 178,902	$ (121,591)
Other comprehensive income (loss):				
Unrealized gain (loss) on short-term investments and currency translation adjustments, net of tax	20	(72)	(4)	(11)
Total other comprehensive income (loss)	20	(72)	(4)	(11)
Total comprehensive income (loss)	$ 224,113	$ (30,616)	$ 178,898	$ (121,602)

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

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

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In	Accumulated Other Comprehensive	Accumulated	Total Shareholders'
	Shares	Amount	Capital	Loss	Deficit	Equity
Balances at December 31, 2021	69,060,403	$ 18	$ 860,482	$ (66)	$ (675,397)	$ 185,037
Issuance of Class A common shares under incentive award plans	210,720	1	422	—	—	423
Share-based compensation expense	—	—	6,031	—	—	6,031
Unrealized loss on short-term investments and currency translation adjustments	—	—	—	(37)	—	(37)
Net loss	—	—	—	—	(25,210)	(25,210)
Balances at March 31, 2022	69,271,123	$ 19	$ 866,935	$ (103)	$ (700,607)	$ 166,244
Issuance of Class A common shares under incentive award plans	155,644	—	542	—	—	542
Share-based compensation expense	—	—	6,676	—	—	6,676
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	13	—	13
Net loss	—	—	—	—	(19,981)	(19,981)
Balances at June 30, 2022	69,426,767	$ 19	$ 874,153	$ (90)	$ (720,588)	$ 153,494
Issuance of Class A common shares under incentive award plans	97,571	—	360	—	—	360
Share-based compensation expense	—	—	6,041	—	—	6,041
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	20	—	20
Net income	—	—	—	—	224,093	224,093
Balances at September 30, 2022	69,524,338	$ 19	$ 880,554	$ (70)	$ (496,495)	$ 384,008

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In	Accumulated Other Comprehensive	Accumulated	Total Shareholders'
	Shares	Amount	Capital	Income	Deficit	Equity
Balances at December 31, 2020	68,215,022	$ 18	$ 829,424	$ (34)	$ (517,473)	$ 311,935
Issuance of Class A common shares under incentive award plans	115,012	—	1,106	—	—	1,106
Share-based compensation expense	—	—	7,126	—	—	7,126
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	13	—	13
Net loss	—	—	—	—	(49,484)	(49,484)
Balances at March 31, 2021	68,330,034	$ 18	$ 837,656	$ (21)	$ (566,957)	$ 270,696
Issuance of Class A common shares under incentive award plans	134,715	—	887	—	—	887
Share-based compensation expense	—	—	5,717	—	—	5,717
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	48	—	48
Net loss	—	—	—	—	(41,563)	(41,563)
Balances at June 30, 2021	68,464,749	$ 18	$ 844,260	$ 27	$ (608,520)	$ 235,785
Issuance of Class A common shares under incentive award plans and employee share purchase plan	423,686	—	2,680	—	—	2,680
Share-based compensation expense	—	—	6,199	—	—	6,199
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	(72)	—	(72)
Net loss	—	—	—	—	(30,544)	(30,544)
Balances at September 30, 2021	68,888,435	$ 18	$ 853,139	$ (45)	$ (639,064)	$ 214,048

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

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

	Nine Months Ended September 30,	
	2022	**2021**
Cash flows from operating activities:		
Net income (loss)	$ 178,902	$ (121,591)
Adjustments to reconcile net income (loss) to net cash used in operating activities:		
Depreciation and amortization expense	1,800	1,687
Share-based compensation expense	18,748	19,042
Non-cash lease expense	2,253	1,895
Amortization of premiums and accretion of discounts on short-term investments	121	617
Loss on disposal of property and equipment	23	—
Deferred income taxes	(185,843)	—
Changes in operating assets and liabilities:		
Prepaid expenses and other current assets	(8,726)	1,034
Accounts receivable, net	(7,174)	(3,224)
Inventory	(10,946)	(5,606)
Other long-term assets	2,919	(1,059)
Accounts payable	(399)	1,116
Accrued expenses and other current liabilities	82	295
Operating lease liabilities	(2,217)	(1,700)
Deferred revenue	26,290	—
Other long-term liabilities	1,550	80
Net cash provided (used) in operating activities	17,383	(107,414)
Cash flows from investing activities:		
Proceeds from sale of property and equipment	81	—
Purchases of property and equipment	(137)	(164)
Purchases of short-term investments	(73,062)	(97,460)
Proceeds from the maturities of short-term investments	107,700	296,300
Intangible asset acquired	—	(20,000)
Net cash provided by investing activities	34,582	178,676
Cash flows from financing activities:		
Proceeds from issuance of Class A common shares under incentive award plans and employee share purchase plan	2,127	4,673
Payments in connection with Common Stock tendered for employee tax obligations	(801)	—
Net cash provided by financing activities	1,326	4,673
Net increase in cash, cash equivalents and restricted cash	53,291	75,935
Cash, cash equivalents and restricted cash at beginning of period	122,470	114,248
Cash, cash equivalents and restricted cash at end of period	$ 175,761	$ 190,183
Supplemental information:		
Cash paid for income taxes	$ 4,608	$ 859
Supplemental disclosure of non-cash investing and financing activities:		
Right-of-use asset obtained in exchange for operating lease obligation	$ 2,876	$ 1,462

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

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

1. Nature of the Business and Basis of Presentation

Kiniksa Pharmaceuticals, Ltd. (the "Company") is a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. The Company's portfolio of assets is based on strong biologic rationale or validated mechanisms, target underserved conditions and offer the potential for differentiation.

The Company is subject to risks and uncertainties common to early, commercial stage companies in the biopharmaceutical industry and global health, societal, economic and market conditions, including the Company's dependence on third parties, including contract research organizations and contract manufacturing organizations, the Company's limited experience obtaining regulatory approvals, the potential failure of the Company to successfully complete research and development of its current or future product candidates, the potential inability of the Company to adequately protect its technology, potential competition, the uncertainty that any current or future product candidates will obtain necessary government regulatory approval, that ARCALYST will continue to be commercially viable, whether any of the Company's current or future product candidates, if approved, will be commercially viable, adverse impact from the coronavirus disease 2019 ("COVID-19") pandemic and global and political turmoil, including the ongoing war in Ukraine. Such risks and uncertainties, especially those risks and uncertainties arising from the ongoing COVID-19 pandemic, may be subject to substantial and uncertain changes, which may cause significant disruption to the Company's business and operations, preclinical studies and clinical trials, the business and operations of the third parties with whom the Company conducts business and the national and global economies, all of which may have material impacts on the Company's business, financial condition and results of operations.

Principles of Consolidation

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

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 recognition of revenue, the capitalization of inventory, the accrual for research and development expenses, the valuation of share-based awards and the realizability of deferred tax assets. 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 information included in this quarterly report on Form 10-Q should be read in conjunction with the Company's audited consolidated financial statements and the accompanying notes included in the Company's Annual Report on Form 10-K for the year

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

ended December 31, 2021 (the "2021 Form 10-K"). The Company's accounting policies are described in the Notes to Consolidated Financial Statements in the Company's 2021 Form 10-K and updated, as necessary, in this report. The accompanying year-end consolidated balance sheet was derived from audited financial statements but does not include all disclosures required by GAAP. The unaudited interim consolidated financial statements have been prepared on the same basis as the audited annual consolidated financial statements and, in the opinion of management, reflect all adjustments, which include only normal recurring adjustments, necessary for the fair statement of the Company's financial position as of September 30, 2022 and the results of its operations for the three and nine months ended September 30, 2022 and 2021, the changes in its shareholders' equity for the three and nine months ended September 30, 2022 and 2021 and its cash flows for the nine months ended September 30, 2022 and 2021. The results for the three and nine months ended September 30, 2022 are not necessarily indicative of results to be expected for the year ending December 31, 2022, any other interim periods or any future year or period.

Liquidity

The Company has evaluated whether there are conditions and events, considered in the aggregate, that raise substantial doubt about the Company's ability to continue as a going concern within one year after the date the consolidated financial statements are issued. As of September 30, 2022, the Company had an accumulated deficit of $496,495. During the nine months ended September 30, 2022, the Company reported net income of $178,902 and provided $17,383 cash in operating activities. As of September 30, 2022, the Company had cash, cash equivalents and short-term investments of $200,724.

Based on its current operating plan, the Company expects that its cash, cash equivalents and short-term investments will be sufficient to fund its operations 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 fund its operations through sales of ARCALYST and/or raise additional capital, as needed. If the Company is unable to grow sales of ARCALYST in future periods, the Company would need to seek additional financing through public or private securities offerings, debt financings, government funding or grants, or other sources, which may include licensing, collaborations or other strategic transactions or arrangements. Although the Company has been successful in raising capital in the past, there is no assurance that it will be successful in obtaining such additional financing on terms acceptable to the Company, if at all. If the Company is unable to obtain funding, the Company could be forced to delay, reduce or eliminate some or all of its commercialization efforts, research and development programs for product candidates or product portfolio expansion, which could adversely affect its business prospects, or the Company may be unable to continue operations.

2. Summary of Significant Accounting Policies

Revenue Recognition

ASC Topic 606, Revenue from Contracts with Customers ("ASC 606") outlines a five-step process for recognizing revenue from contracts with customers: (i) identify the contract with the customer, (ii) identify the performance obligations in the contract, (iii) determine the transaction price, (iv) allocate the transaction price to the separate performance obligations in the contract, and (v) recognize revenue associated with the performance obligations as they are satisfied.

The Company only applies the five-step model to contracts when it is probable that the Company will collect the consideration it is entitled to in exchange for the goods or services it transfers to the customer. Once a contract is determined to be within the scope of ASC 606, the Company determines the performance obligations that are distinct. The Company recognizes as revenues the amount of the transaction price that is allocated to each respective performance obligation when the performance obligation is satisfied. Generally, the Company's performance obligations are transferred to customers at a point in time, typically upon delivery of the product to the customer.

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ASC 606 requires entities to record a contract asset when a performance obligation has been satisfied or partially satisfied, but the amount of consideration has not yet been received because the receipt of the consideration is conditioned on something other than the passage of time. ASC 606 also requires an entity to present a revenue contract as a contract liability in instances when a customer pays consideration, or an entity has a right to an amount of consideration that is unconditional (e.g., receivable), before the entity transfers a good or service to the customer.

Collaboration Revenue

The Company analyzes its collaboration arrangements to assess whether such arrangements involve joint operating activities performed by parties that are both active participants in the activities and exposed to significant risks and rewards dependent on the commercial success of such activities and therefore within the scope of ASC Topic 808, Collaborative Arrangements ("Topic 808"). This assessment is performed throughout the life of the arrangement based on changes in the responsibilities of all parties in the arrangement. For collaboration arrangements within the scope of Topic 808 that contain multiple elements, the Company first determines which elements of the collaboration are deemed to be within the scope of Topic 808 and which elements of the collaboration are more reflective of a vendor-customer relationship and therefore within the scope of Topic 606.

For elements of collaboration arrangements that are accounted for pursuant to ASC 606, we identify the performance obligations and allocate the total consideration we expect to receive on a relative standalone selling price basis to each performance obligation. Variable consideration such as performance-based milestones will be included in the total consideration if we expect to receive such consideration and if it is probable that the inclusion of the variable consideration will not result in a significant reversal in the cumulative amount of revenue recognized under the arrangement. Our estimate of the total consideration we expect to receive under each collaboration arrangement is updated for each reporting period, and any adjustments to revenue are recorded on a cumulative catch-up basis. We exclude sales-based royalty and milestone payments from the total consideration we expect to receive until the underlying sales occur because the license to our intellectual property is deemed to be the predominant item to which the royalties or milestones relate as it is the primary driver of value in our collaboration arrangements.

Key assumptions to determine the standalone selling price may include forecasted revenues, development timelines, reimbursement rates for personnel costs, discount rates and probabilities of technical and regulatory success. We recognize revenue associated with each performance obligation as the control over the promised goods or services transfer to our collaboration partner which occurs either at a point in time or over time. If control transfers over time, revenue is recognized by using a method of measuring progress that best depicts the transfer of goods or services. We evaluate the measure of progress and related inputs each reporting period and any resulting adjustments to revenue are recorded on a cumulative catch-up basis.

Consideration received that does not meet the requirements to satisfy ASC 808 or ASC 606 revenue recognition criteria is recorded as deferred revenue in the accompanying consolidated balance sheets, classified as either short-term (less than 12 months) or long-term (more than 12 months) deferred revenue based on our best estimate of when such revenue will be recognized.

There have been no other material changes to the significant accounting policies previously disclosed in the Company's 2021 Form 10-K.

Recently Adopted Accounting Pronouncements

Accounting standards that have been issued by the Financial Accounting Standards Board or other standards-setting bodies that do not require adoption until a future date are not expected to have a material impact on the Company's financial statements upon adoption.

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3. Fair Value of Financial Assets and Liabilities

Financial assets and liabilities carried at fair value are to be classified and disclosed in one of the following three levels of the fair value hierarchy, of which the first two are considered observable and the last is considered unobservable:

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

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

	Fair Value Measurements as of September 30, 2022 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Cash equivalents — money market funds	$ 39,533	$ —	$ —	$ 39,533
Short-term investments — U.S. Treasury notes	—	24,963	—	24,963
	$ 39,533	$ 24,963	$ —	$ 64,496

	Fair Value Measurements as of December 31, 2021 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Cash equivalents — money market funds	$ 94,324	$ —	$ —	$ 94,324
Short-term investments — U.S. Treasury notes	—	59,731	—	59,731
	$ 94,324	$ 59,731	$ —	$ 154,055

During the nine months ended September 30, 2022 and the year ended December 31, 2021 there were no transfers between Level 1, Level 2 and Level 3. The money market funds were valued using quoted prices in active markets, which represent a Level 1 measurement in the fair value hierarchy. The Company's cash equivalents and short-term investments as of September 30, 2022 and December 31, 2021 included U.S. Treasury notes, which are not traded on a daily basis and, therefore, represent a Level 2 measurement in the fair value hierarchy at each period end.

	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Credit Losses	Fair Value
September 30, 2022					
Short-term investments — U.S. Treasury notes	$ 24,986	$ —	$ (23)	$ —	$ 24,963
	$ 24,986	$ —	$ (23)	$ —	$ 24,963

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	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Credit Losses	Fair Value
December 31, 2021					
Short-term investments — U.S. Treasury notes	$ 59,745	$ 1	$ (15)	$ —	$ 59,731
	$ 59,745	$ 1	$ (15)	$ —	$ 59,731

As of September 30, 2022, the Company held 6 securities that were in an unrealized loss position. The aggregate fair value of securities held by the Company in an unrealized loss position was $22,466 at September 30, 2022. As of December 31, 2021, the Company held 11 securities that were in an unrealized loss position. The aggregate fair value of securities held by the Company in an unrealized loss position was $49,739 at December 31, 2021. As of September 30, 2022 and December 31, 2021, these securities were held by the Company in an unrealized loss position for less than 12 months. The Company determined that there was no material change in the credit risk of these securities. As a result, the Company determined it did not hold any investments with an other-than-temporary impairment as of September 30, 2022 and December 31, 2021.

4. Product Revenue, Net

ARCALYST

Following the approval by the U.S. Food and Drug Administration ("FDA") of ARCALYST on March 18, 2021, the Company began generating product revenue from sales of ARCALYST in April 2021.

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2022	2021	2022	2021
Product revenue, net	$ 33,424	$ 12,095	$ 82,585	$ 19,799

The following table summarizes balances and activity in each of the product revenue allowance and reserve categories for the nine months ended September 30, 2022:

	Contractual Adjustments	Government Rebates	Returns	Total
Balance at December 31, 2021	$ 515	$ 719	$ 101	$ 1,335
Current provisions relating to sales in the current year	4,993	3,011	227	8,231
Payments/returns relating to sales in the current year	(3,797)	(1,659)	—	(5,456)
Payments/returns relating to sales in the prior years	(295)	(535)	—	(830)
Balance at September 30, 2022	$ 1,416	$ 1,536	$ 328	$ 3,280

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Total revenue-related reserves as of September 30, 2022 and December 31, 2021, included in our consolidated balance sheets, are summarized as follows:

	September 30, 2022	December 31, 2021
Reduction of accounts receivable	$ (157)	$ (50)
Components of other current liabilities	3,437	1385
Total revenue-related reserves	$ 3,280	$ 1,335

5. Inventory

Inventory consisted of the following:

	September 30, 2022	December 31, 2021
Raw materials	$ —	$ —
Work-in-process	—	—
Finished Goods	14,621	3,675
	$ 14,621	$ 3,675

6. Property and Equipment, Net

Property and equipment, net consisted of the following:

	September 30, 2022	December 31, 2021
Furniture, fixtures and vehicles	$ 224	$ 62
Computer hardware and software	345	341
Leasehold improvements	3,931	3,931
Lab equipment	4,068	4,249
Construction in progress	32	166
Total property and equipment	8,600	8,749
Less: Accumulated depreciation	(6,605)	(5,915)
Total property and equipment, net	$ 1,995	$ 2,834

Depreciation expense was $177 and $373 during the three months ended September 30, 2022 and 2021, respectively, and $885 and $1,079 during the nine months ended September 30, 2022 and 2021, respectively.

7. Intangible Assets

Intangible assets, net of accumulated amortization, impairment charges and adjustments as of September 30, 2022 and December 31, 2021 are summarized in the following table.

	Estimated life	As of September 30, 2022			As of December 31, 2021		
		Cost	Accumulated Amortization	Net	Cost	Accumulated Amortization	Net
Regulatory milestone	20 years	$ 20,000	$ 1,500	$ 18,500	$ 20,000	$ 750	$ 19,250
		$ 20,000	$ 1,500	$ 18,500	$ 20,000	$ 750	$ 19,250

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8. Accrued Expenses

Accrued expenses consisted of the following:

	September 30, 2022	December 31, 2021
Accrued research and development expenses	$ 13,559	$ 24,977
Accrued employee compensation and benefits	8,316	8,916
Accrued collaboration expenses	4,660	835
Accrued legal, commercial and professional fees	3,754	2,933
Other	579	370
	$ 30,868	$ 38,031

9. Share-Based Compensation

The Company maintains several equity compensation plans, including the 2018 Incentive Award Plan (the "2018 Plan"), 2018 Employee Share Purchase Plan (the "2018 ESPP"), and Rilonacept Long-Term Incentive Plan ("RLTIP") which was approved under the 2018 Plan. Upon the effectiveness of the 2018 Plan, the Company ceased granting awards under its 2015 Equity Incentive Plan (as amended, the "2015 Plan" and together with the 2018 Plan, the "Plans").

2015 Plan

As of September 30, 2022, there were 2,029,355 Class A common shares subject to outstanding awards under the 2015 Plan and reserved for issuance thereunder pursuant to such awards.

2018 Plan

In May 2018, the Company's board of directors and shareholders approved the 2018 Plan, which became effective on May 23, 2018. The 2018 Plan provides for the grant of incentive share options, nonqualified share options, share appreciation rights, restricted shares, dividend equivalents, restricted share units ("RSUs") and other share- or cash- based awards. Pursuant to the 2018 Plan's evergreen provision, the number of shares available for future issuance under the 2018 Plan, as of January 1, 2022, increased by 2,762,416 Class A common shares. As of September 30, 2022, 4,117,546 shares remained available for future grant under the 2018 Plan.

2018 ESPP

In December 2021, pursuant to the 2018 ESPP's evergreen provision, the Company's board of directors approved an increase in the number of shares available for future issuance under the 2018 ESPP, as of January 1, 2022, of 90,000 Class A common shares. As of September 30, 2022, 604,290 Class A common shares were available for future issuance under the 2018 ESPP.

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Options

Share option activity under the Plans is summarized as follows:

	Number of Shares	Weighted Average Exercise Price
Outstanding as of December 31, 2021	9,226,846	$ 14.14
Granted	2,792,460	$ 11.22
Exercised	(226,230)	$ 5.81
Forfeited	(1,468,620)	$ 15.59
Outstanding as of September 30, 2022	10,324,456	$ 13.32
Share options exercisable as of September 30, 2022	5,525,898	$ 13.52
Share options unvested as of September 30, 2022	4,798,558	$ 13.10

As of September 30, 2022, total unrecognized compensation expense related to the unvested share option awards was $40,360 which is expected to be recognized over a weighted average remaining period of 2.73 years.

Restricted Share Units

Beginning in March 2021, the Company began granting RSUs with service conditions (“Time-Based RSUs”) to eligible employees as part of its equity incentive compensation. The Time-Based RSUs vest 25% on each of the first, second, third and fourth anniversaries of the date of grant, subject to continued employment through such dates.

During the years ended December 31, 2020 and 2019, the Company granted the first RSU awards (“First RLTIP RSU Awards”) as part of the RLTIP to eligible employees. During the year ended December 31, 2021, the FDA Milestone (as defined in RLTIP) was achieved (the date of such achievement, the “Achievement Date”) and (1) the number of Class A common shares issuable under the First RLTIP RSU Awards were determined in accordance with the RLTIP and vested in one installment on the first anniversary of the Achievement Date, subject to continued employment through such date, and (2) the Company granted a second set of RSU awards to eligible employees on the Achievement Date with respect to a number of shares determined in accordance with the RLTIP, which will vest on the second anniversary of the Achievement Date, subject to continued employment through such date.

During the three months ended September 30, 2022 and 2021, the Company recognized compensation expense of $1,146 and $838, respectively, related to RSUs including those granted in connection with the RLTIP. During the nine months ended September 30, 2022 and 2021, the Company recognized compensation expense of $2,697 and $2,868, respectively, related to RSUs including those granted in connection with the RLTIP.

The following table summarizes RSU activity, including the RSUs outstanding under the RLTIP for the nine months ended September 30, 2022:

	Number of Shares	Weighted Average Grant Date Fair Value
Unvested RSUs as of December 31, 2021	885,021	$ 15.72
Granted	1,318,583	$ 10.71
Vested	(229,151)	$ 15.79
Forfeited	(320,710)	$ 14.08
Unvested RSUs as of September 30, 2022	1,653,743	$ 13.17

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As of September 30, 2022, total unrecognized compensation cost related to the RSU awards was $17,821 which is expected to be recognized over a weighted average remaining period of 3.38 years.

Share-Based Compensation

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

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2022	2021	2022	2021
Cost of goods sold	$ 164	$ 62	$ 456	$ 101
Research and development expenses	1,532	1,912	5,116	6,469
Selling, general and administrative expenses	4,345	4,225	13,176	12,472
	$ 6,041	$ 6,199	$ 18,748	$ 19,042

10. Out-Licensing Agreements

Genentech License Agreement

On August 2, 2022, the Company entered into a license agreement (the "Genentech License Agreement") with Genentech, Inc. and F. Hoffmann-La Roche Ltd (collectively, "Genentech"), pursuant to which the Company granted Genentech exclusive worldwide rights to develop, manufacture and commercialize vixarelimab and related antibodies (each, a "Genentech Licensed Product"). The Genentech License Agreement became effective on September 12, 2022 (the "Genentech Effective Date") following termination of the statutory waiting period under the Hart-Scott Rodino Act.

Under the Genentech License Agreement, the Company received an upfront payment of $80,000 for the license and is eligible to receive a near-term cash payment of $20,000 for delivery of certain materials to Genentech. In addition, the Company will be eligible to receive up to approximately $600,000 in contingent payments, including specified development, regulatory and sales-based milestones, before fulfilling the Company's upstream financial obligations. The Company will also be eligible to receive tiered percentage royalties on a Genentech Licensed Product-by-Genentech Licensed Product basis ranging from low-double digits to mid-teens on annual net sales of each Genentech Licensed Product, subject to certain customary reductions, with an aggregate minimum floor, before fulfilling the Company's upstream financial obligations. Royalties will be payable on a Genentech Licensed Product-by-Genentech Licensed Product and country-by-country basis until the latest to occur of the expiration of certain patents that cover a Genentech Licensed Product, the expiration of regulatory exclusivity for such Genentech Licensed Product, or the tenth anniversary of first commercial sale of such Genentech Licensed Product in such country.

Pursuant and subject to the terms of the Genentech License Agreement, Genentech has the exclusive worldwide right to conduct development and commercialization activities for Genentech Licensed Products at its sole cost. Notwithstanding the foregoing, the Company is responsible, at its sole cost, for continuing to conduct and finalize its Phase 2b clinical trial assessing the efficacy, safety and tolerability of vixarelimab in reducing pruritis in prurigo nodularis. Both the Company and Genentech participate in a joint transition committee, which coordinates and oversees the technology and inventory transition activities relating to the development of the Genentech Licensed Products and the Company's conduct and finalization of its Phase 2b clinical trial.

The Company will provide to Genentech an initial drug supply and a drug product resupply and Genentech will be obligated to pay $20,000 to the Company after delivery of the last of such supply; *provided* that the materials are delivered by the applicable required supply date and meet the specified requirements. Genentech has the right, for a specified period of time, to purchase an additional batch of vixarelimab drug substance under the Genentech License

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Agreement at cost plus a markup. Under the Genentech License Agreement, Genentech has the right to assume manufacturing responsibilities for Genentech Licensed Products.

Absent early termination, the Genentech License Agreement will continue until there are no more royalty or other payment obligations owed to the Company. Genentech has the right to terminate the Genentech License Agreement at its discretion with prior written notice and either party may terminate the Genentech License Agreement in the event of an uncured material breach of the other party or in the case of insolvency of the other party. In addition, the Genentech License Agreement will terminate upon termination of the Biogen Agreement (as defined below).

The Company concluded that Genentech is a customer in this license agreement, and as such, the Genentech License Agreement falls within the scope of the revenue recognition guidance in ASC 606.

Accounting for Genentech License Agreement

As of the Genentech Effective Date, the Company identified the following material promises in the Genentech License Agreement: (i) the delivery of the exclusive license for vixarelimab; (ii) an initial drug supply delivery; (iii) a drug product resupply delivery; and (iv) completion of the Phase 2b clinical trial for vixarelimab.

The Company also evaluated whether certain options outlined within the Genentech License Agreement represented material rights that would give rise to a performance obligation, including the option to purchase additional drug substance, and concluded that none of the options convey a material right to Genentech and therefore are not considered separate performance obligations within the Genentech License Agreement.

The Company assessed the above promises and determined that the exclusive license for vixarelimab is reflective of a vendor-customer relationship and therefore represents a performance obligation. The exclusive license for vixarelimab is considered functional intellectual property and distinct from other promises under the Genentech License Agreement as Genentech can benefit from the license on its own or together with other readily available resources and the license is separately identifiable from the other promises. The initial drug supply and drug product resupply are considered distinct from the exclusive license for vixarelimab as Genentech can benefit from such supply together with the license transferred by the Company at the inception of the Genentech License Agreement. The completion of the Phase 2b clinical trial is considered distinct from the exclusive license for vixarelimab as Genentech can benefit from the data generated by such trial together with such license. Therefore, each represents a separate performance obligation within a contract with a customer at contract inception.

The Company determined the transaction price at the inception of the Genentech License Agreement which consists of the $80,000 upfront payment. The Company determined that any variable consideration, including the $20,000 related to the delivery of the initial drug supply and drug product resupply and future development and regulatory milestones, are deemed fully constrained and therefore excluded from the transaction price due to the high degree of uncertainty and risk associated with these potential payments, as the Company also determined that it could not assert that it was not probable that a significant reversal in the amount of cumulative revenue recognized would occur. The Company also determined that royalties and sales milestones relate solely to the license of intellectual property. Revenue related to these royalties and sales milestones will only be recognized when the associated sales occur, and relevant thresholds are met, under the sales or usage-based royalty exception of Topic 606.

As noted above, the Company identified four performance obligations in the Genentech License Agreement: (i) the delivery of the exclusive license for vixarelimab; (ii) an initial drug supply delivery; (iii) a drug product resupply delivery; and (iv) completion of the Phase 2b clinical trial for vixarelimab. The selling price of each performance obligation in the Genentech License Agreement was determined based on the Company's standalone selling price ("SSP") with the objective of determining the price at which it would sell such an item if it were to be sold regularly on a standalone basis. The Company allocated the transaction price to the exclusive license, initial drug supply delivery, drug product resupply delivery and completion of the Phase 2b clinical trial for vixarelimab. The Company recognizes

revenue for the license performance obligations at a point in time, that is upon transfer of the license to Genentech. As control of the license was transferred on the Genentech Effective Date and Genentech could begin to use and benefit from the license, the Company recognized $65,711 of collaboration revenue during the three and nine months ended September 30, 2022 under the Genentech License Agreement. The Company will recognize revenue for deliveries of certain drug supply to Genentech at a point in time upon delivery. The Company will recognize revenue for the completion of the Phase 2b clinical trial for vixarelimab over time using the cost-to-cost input method, which it believes best depicts the transfer of control to the customer. Under the cost-to-cost input method, the extent of progress towards completion is measured based on the ratio of actual costs incurred to the total estimated costs expected upon satisfying the identified performance obligation. Under this method, revenue is recorded as a percentage of the allocated transaction price based on the progress of completion. The calculation of the total estimated costs to fulfill the performance obligation includes costs associated with employees, development, manufacturing, and out-of-pocket costs expected to be paid to third parties.

Huadong Collaboration Agreements

On February 21, 2022 (the "Effective Date"), the Company entered into two collaboration and license agreements (each, a "Collaboration Agreement" and together, the "Collaboration Agreements") with Hangzhou Zhongmei Huadong Pharmaceutical Co., Ltd. ("Huadong"), pursuant to which the Company granted Huadong exclusive rights to develop and commercialize rilonacept and develop, manufacture and commercialize mavrilimumab (each, a "Licensed Product" and together, the "Licensed Products") in the following countries: People's Republic of China, Hong Kong SAR, Macao SAR, Taiwan Region, South Korea, Indonesia, Singapore, The Philippines, Thailand, Australia, Bangladesh, Bhutan, Brunei, Burma, Cambodia, India, Laos, Malaysia, Maldives, Mongolia, Nepal, New Zealand, Sri Lanka, and Vietnam (collectively, the "Territory"). The Company otherwise retained its current rights to the Licensed Products outside the Territory.

Under the Collaboration Agreements, the Company received a total upfront cash payment of $22,000, which includes $12,000 for the Territory license of rilonacept and $10,000 for the Territory license of mavrilimumab. The Company will be eligible to receive up to approximately $70,000 in payments for rilonacept, and up to approximately $576,000 in payments for mavrilimumab, including specified development, regulatory and sales-based milestones. Huadong will also be obligated to pay the Company tiered percentage royalties on a Licensed Product-by-Licensed Product basis ranging from the low-teens to low-twenties on annual net sales of each Licensed Product in the Territory, subject to certain reductions tied to rilonacept manufacturing costs and certain other customary reductions, with an aggregate minimum floor. Royalties will be payable on a Licensed Product-by-Licensed Product and country-by-country or region-by-region basis until the later of (i) 12 years after the first commercial sale of the applicable Licensed Product in such country or region in the Territory, (ii) the date of expiration of the last valid patent claim of the Company's patent rights or any joint collaboration patent rights that covers the applicable Licensed Product in such country or region in the Territory, and (iii) the expiration of the last regulatory exclusivity for the applicable Licensed Product in such country or region in the Territory.

Pursuant and subject to the terms of the Collaboration Agreements, Huadong has the exclusive right to conduct Territory-specific development activities for the Licensed Products in the Territory, the first right to support global development of the Licensed Products by serving as the sponsor of the global clinical trials conducted in the Territory and the exclusive right to commercialize the Licensed Products in the Territory. Huadong will be responsible for all costs of development activities and commercialization in the Territory. Both the Company and Huadong participate in a Joint Steering Committee ("JSC"), which coordinates and oversees the exploitation of the Licensed Products in the Territory.

The Company will supply certain materials to support development and commercialization activities for both mavrilimumab and rilonacept. Under the Collaboration Agreement for mavrilimumab, Huadong has the right to assume manufacturing responsibilities for materials in the Territory. Under the Collaboration Agreement for rilonacept, Huadong does not have rights to perform manufacturing activities in the Territory.

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Absent early termination, each Collaboration Agreement will continue on a country-by-country or region-by-region basis until there are no more royalty payments owed to the Company in such country or region for the applicable Licensed Product. Huadong has the right to terminate each Collaboration Agreement at its discretion upon 12 months' notice and either party may terminate the applicable Collaboration Agreement in the event of an uncured material breach of the other party or in the case of insolvency of the other party. In addition, the Company may terminate the applicable Collaboration Agreement if Huadong or its affiliates or sublicensees challenges the scope, validity, or enforceability of the Company's patent rights being licensed to Huadong. If Huadong and its affiliates do not conduct any material development or commercialization activities with respect to a Licensed Product in the People's Republic of China for a continuous period of longer than six months, then, subject to certain exceptions, the Company may terminate the Collaboration Agreement applicable to such Licensed Product with 60 days' prior written notice. In addition, Huadong's rights under each Collaboration Agreement in certain regions within the Territory may be subject to termination upon failure by Huadong to perform certain clinical, development or commercialization activities, as applicable, with respect to the applicable Licensed Product in such regions.

The Company concluded that Huadong is a customer in these Collaboration Agreements, and as such, each Collaboration Agreement falls within the scope of the revenue recognition guidance in ASC 606.

The Company concluded that the Collaboration Agreements should not be combined and treated as a single arrangement for accounting purposes as the Collaboration Agreements were negotiated separately with separate and distinct commercial objectives, the amount of consideration in one Collaboration Agreement is not dependent on the price or performance of the other Collaboration Agreement, and the goods and services promised in the Collaboration Agreements are not a single performance obligation.

Accounting for Mavrilimumab Collaboration Agreement

As of the Effective Date, the Company identified the following material promises in the mavrilimumab Collaboration Agreement: delivery of (i) exclusive license for mavrilimumab in the Territory and (ii) clinical manufacturing supply of certain materials for mavrilimumab products in the Territory.

The Company also evaluated whether certain options outlined within the mavrilimumab Collaboration Agreement represented material rights that would give rise to a performance obligation and concluded that none of the options convey a material right to Huadong and therefore are not considered separate performance obligations within the mavrilimumab Collaboration Agreement.

The Company assessed the above promises and determined that the exclusive license for mavrilimumab in the Territory is reflective of a vendor-customer relationship and therefore represents a performance obligation. The exclusive license for mavrilimumab in the Territory is considered functional intellectual property and distinct from other promises under the Collaboration Agreement as Huadong can benefit from the license on its own or together with other readily available resources and the license is separately identifiable from the other promises. The clinical manufacturing supply of certain materials for mavrilimumab products in the Territory is considered distinct from the exclusive license for mavrilimumab as Huadong can benefit from the manufacturing services together with the license transferred by the Company at the inception of the Collaboration Agreement. Therefore, each represents a separate performance obligation within a contract with a customer at contract inception.

The Company determined the transaction price at the inception of the mavrilimumab Collaboration Agreement which includes $10,000, consisting of the upfront payment. The Company also includes an estimate of variable consideration associated with the clinical manufacturing supply of certain materials when those materials are shipped. The Company determined that any variable consideration related to development and regulatory milestones is deemed fully constrained and therefore excluded from the transaction price due to the high degree of uncertainty and risk associated with these potential payments, as the Company determined that it could not assert that it was probable that a significant reversal in the amount of cumulative revenue recognized will not occur. The Company also determined that

royalties and sales milestones relate solely to the licenses of intellectual property. Revenue related to these royalties and sales milestones will only be recognized when the associated sales occur, and relevant thresholds are met, under the sales or usage-based royalty exception of Topic 606.

As noted above, the Company identified two performance obligations in the mavrilimumab Collaboration Agreement: (i) the delivery of the exclusive license for mavrilimumab in the Territory; and (ii) the clinical manufacturing supply of certain materials for mavrilimumab products in the Territory. The selling price of each performance obligation in the mavrilimumab Collaboration Agreement was determined based on the Company's standalone selling price ("SSP") with the objective of determining the price at which it would sell such an item if it were to be sold regularly on a standalone basis. The Company allocated the variable consideration related to the manufacturing obligations to the future clinical supply of mavrilimumab products in the Territory and the remaining fixed and variable consideration to the license obligation. The Company recognizes revenue for the license performance obligations at a point in time, that is upon transfer of the license to Huadong. As control of the license was transferred on the Effective Date and Huadong could begin to use and benefit from the license, the Company recognized $10,000 of collaboration revenue during the nine months ended September 30, 2022 under the mavrilimumab Collaboration Agreement. The Company will recognize revenue for the clinical manufacturing supply obligations at a point in time, that is upon each delivery of the supply to Huadong.

Accounting for Rilonacept Collaboration Agreement

As of the Effective Date, the Company identified the following material promises in the rilonacept Collaboration Agreement that were evaluated: delivery of (i) exclusive license for rilonacept in the Territory; (ii) clinical manufacturing supply of certain materials for rilonacept products in the Territory; and (iii) commercial manufacturing supply of certain material for rilonacept products in the Territory.

The Company also evaluated whether certain options outlined within the rilonacept Collaboration Agreement represented material rights that would give rise to a performance obligation and concluded that none of the options convey a material right to Huadong and therefore are not considered separate performance obligations within the rilonacept Collaboration Agreement.

The Company assessed the above promises and determined that there is one combined performance obligation for the exclusive license for rilonacept and clinical and commercial manufacturing obligations for rilonacept products in the Territory. Huadong cannot exploit the value of the exclusive license for rilonacept products in the Territory without receipt of supply as the exclusive license for rilonacept products in the Territory does not convey to Huadong the right to manufacture and therefore the Company has combined the exclusive license for rilonacept products in the Territory and the manufacturing obligations into one performance obligation.

The Company determined the transaction price at the inception of the rilonacept Collaboration Agreement which includes $12,000, consisting of the upfront payment. The Company also includes an estimate of variable consideration associated with the clinical manufacturing supply of certain materials when those materials are shipped. The Company determined that any variable consideration related to development and regulatory milestones, sales milestones and royalties are deemed fully constrained and therefore excluded from the transaction price due to the high degree of uncertainty and risk associated with these potential payments, as the Company determined that it could not assert that it was probable that a significant reversal in the amount of cumulative revenue recognized will not occur. Royalties and sales milestones will be recognized as the Company delivers the commercial manufactured product to Huadong. Any changes in estimates may result in a cumulative catch-up based on the number of units of manufactured product delivered.

As noted above, the Company identified a single combined performance obligation in the rilonacept Collaboration Agreement consisting of the exclusive license for rilonacept and clinical and commercial manufacturing obligations for rilonacept products in the Territory. The Company recognizes revenue for the combined performance obligation

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

consisting of the exclusive license for rilonacept and clinical and commercial manufacturing obligations for rilonacept products in the Territory at a point in time, upon which control of materials are transferred to Huadong for each delivery of the associated materials. The Company currently expects to recognize the revenue over the life of the agreement. This estimate considers the timing of development and commercial activities under the rilonacept Collaboration Agreement and may be reduced or increased based on changes in the various activities.

The Company has not recognized any revenue under the rilonacept Collaboration Agreement for the three and nine months ended September 30, 2022 as there has been no delivery of materials under the rilonacept Collaboration Agreement to date. The full transaction price of $12,000 is recorded in long-term deferred revenue, based upon timing of anticipated future shipments.

The following table summarizes deferred revenue in connection with license and collaboration agreements for the nine months ended September 30, 2022:

	Balance at Beginning of Period	Additions	Deductions	Balance at End of Period
Nine Months Ended September 30, 2022				
Huadong rilonacept	$ —	$ 12,000	$ —	$ 12,000
Genentech vixarelimab	—	14,290	—	14,290
Deferred revenue	$ —	$ 26,290	$ —	$ 26,290

11. In-license, Acquisition and Collaboration Agreements

Biogen Asset Purchase Agreement

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

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, of which $165,000 remains as of September 30, 2022. Additionally, the Company could be obligated to make up to an aggregate of up to $150,000 of payments upon the achievement of specified annual net sales milestones and to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

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

The Biogen Agreement will terminate upon the expiration of all payment obligations with respect to the last product in all countries in the territory. The Company has the right to terminate the agreement with 90 days' prior written notice. Both parties may terminate by mutual written consent or in the event of material breach of the agreement by the other party that remains uncured for 90 days (or 30 days for payment-related breaches).

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In July 2017, the Company and Biogen entered into Amendment No. 1 to the Biogen Agreement, which clarified the scope of the antibodies subject to the Biogen Agreement.

On August 2, 2022, the Company entered into Amendment No. 2 to the Biogen Agreement (the "Second Biogen Amendment"). Pursuant to the terms of the Second Biogen Amendment, commencing on the effective date of the Genentech License Agreement, certain defined terms in the Biogen Agreement were amended, including "Net Sales", "Indication", "Product", "Combination Product" and "Valid Claim". In addition, the tiered royalty rates to be paid by the Company to Biogen increased by an amount equal to less than one percent.

Upon the termination or expiration of the Genentech License Agreement, the amendments to the terms of the Biogen Agreement, as set forth in the Second Biogen Amendment, will terminate and all terms of the Biogen Agreement will revert to the version of such terms in effect as of immediately prior to the effective date of the Genentech License Agreement.

During the three and nine months ended September 30, 2022, the Company recorded research and development expense of $11 and $45, respectively, related to the annual maintenance in connection with the Biogen Agreement. During the three and nine months ended September 30, 2021, the Company recorded research and development expense of $14 and $42 respectively, related to the annual maintenance fee in connection with the retained contracts.

Beth Israel Deaconess Medical Center License Agreement

In 2019, the Company exercised the call option under the stock purchase option agreement with Primatope and acquired all of the outstanding securities of Primatope (the "Primatope Acquisition"). As a result of the Primatope Acquisition, the Company acquired the rights to an exclusive license to certain intellectual property rights controlled by Beth Israel Deaconess Medical Center, Inc. ("BIDMC") to make, use, develop and commercialize KPL-404 (the "BIDMC Agreement"). Under the BIDMC Agreement, the Company is solely responsible for all development, regulatory and commercial activities and costs. The Company is also responsible for costs related to filing, prosecuting and maintaining the licensed patent rights. Under the BIDMC Agreement, the Company is obligated to pay an insignificant annual maintenance fee as well as clinical and regulatory milestone payments of up to an aggregate of $1,200 to BIDMC. The Company is also obligated to pay a low single-digit royalty on annual net sales of products licensed under the agreement.

During the three and nine months ended September 30, 2022 and 2021, the Company did not record any research and development expense in connection with the BIDMC Agreement.

Regeneron License Agreement

In September 2017, the Company entered into a license agreement (the "Regeneron Agreement") with Regeneron Pharmaceuticals, Inc. ("Regeneron"), pursuant to which the Company has been granted an exclusive license under certain intellectual property rights controlled by Regeneron to develop and commercialize ARCALYST worldwide, excluding the Middle East and North Africa, for all indications other than those in oncology and local administration to the eye or ear. Upon receiving positive data in RHAPSODY, the Company's pivotal Phase 3 clinical trial of ARCALYST, Regeneron transferred the biologics license application, for ARCALYST to the Company. In March 2021, when the FDA granted approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older, the Company assumed the sales and distribution of ARCALYST for Cryopyrin-Associated Periodic Syndromes and Deficiency of Interleukin-1 Receptor Antagonist in the United States.

The Company made a $20,000 payment in the first quarter of 2021 in connection with the achievement of a specified regulatory milestone event. The $20,000 milestone was accounted for as an intangible asset and will be

amortized over the life of the underlying asset. Related amortization expense will be recorded as cost of goods sold in the Company's consolidated statement of operations and comprehensive loss.

The Company evenly splits profits on sales of ARCALYST with Regeneron, where profits are determined after deducting from net sales of ARCALYST certain costs related to the manufacturing and commercialization of ARCALYST. Such costs include but are not limited to (i) the Company's cost of goods sold for product used, sold or otherwise distributed for patient use by the Company; (ii) customary commercialization expenses, including the cost of the Company's field force, and (iii) the Company's cost to market, advertise and otherwise promote ARCALYST, with such costs identified in subsection (iii) subject to specified limits. In addition, should there be a transfer of technology related to the manufacture of ARCALYST, then, to the extent permitted in accordance with the Regeneron Agreement, the fully-burdened costs incurred by each of the Company and Regeneron in performing (or having performed) such technology transfer shall also be deducted from net sales of ARCALYST to determine profit. The Company also evenly splits with Regeneron any proceeds received by the Company from any licensees, sublicensees and distributors in consideration for the sale, license or other disposition of rights with respect to ARCALYST, including upfront payments, milestone payments and royalties. For the three and nine months ended September 30, 2022, the Company recognized $4,623 and $16,549 respectively, of expenses related to the profit sharing agreement presented within collaboration expenses. The Company recorded $6,000 of profit sharing expense in February 2022 related to the rilonacept Collaboration Agreement. For the three and nine months ended September 30, 2021 the Company did not recognize any collaboration expenses related to the profit sharing agreement.

Pursuant to the Regeneron Agreement, in September 2017, the parties entered into a clinical supply agreement under which Regeneron agreed to manufacture materials solely for the Company's use in development activities. Pursuant to the Regeneron Agreement, during the year ended December 31, 2021, the Company entered into a commercial supply agreement under which Regeneron agreed to manufacture product for the Company's use, including for commercial sales. The commercial supply agreement terminates upon the termination of the Regeneron Agreement or the date of completion of the transfer of technology related to the manufacture of ARCALYST. During the three and nine months ended September 30, 2022 and 2021, the Company did not incur any research and development expense related to the purchase of drug materials under the clinical supply agreement. As of September 30, 2022 and December 31, 2021, the Company recorded inventory of $14,621 and $3,675, respectively, related to the purchase of commercial product under the commercial supply agreement (see Note 5). As of September 30, 2022, the Company had non-cancelable purchase commitments under the commercial supply agreement (see Note 14).

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 with one year's written notice. The Company may also terminate the agreement with three months' written notice if the licensed product is determined to have certain safety concerns.

MedImmune License Agreement

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

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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, of which $57,500 remain as of September 30, 2022. In addition, the Company is obligated to make clinical and regulatory milestone payments of up to $15,000 in the aggregate for each subsequent indication. In July 2020, the Company entered into an amendment to the MedImmune Agreement to establish a new coronavirus field and defer the payment of certain development and regulatory milestones as applied to the new coronavirus field. The Company is obligated to make milestone payments to MedImmune of up to $85,000 upon the achievement of annual net sales thresholds up to, but excluding, $1,000,000 in annual net sales as well as additional milestone payments aggregating up to $1,100,000 upon the achievement of additional specified annual net sales thresholds starting at $1,000,000 and higher. The Company has also agreed to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the low double-digit percentages and ending at twenty percent. Royalty rates are subject to reductions upon certain events.

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

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

During the three and nine months ended September 30, 2022 and 2021, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

12. Earnings (Loss) per Share

The rights, including the liquidation and dividend rights, of the holders of Class A, Class B, Class A1 and Class B1 common shares are identical, except with respect to voting, transferability and conversion (see the Notes to Consolidated Financial Statements to our Form 10-K). As the liquidation and dividend rights are identical, income and losses are allocated on a proportionate basis, and the resulting net earnings (loss) per share (EPS) attributed to common shareholders will, therefore, be the same for the Class A, A1, B and B1 common shares on an individual or combined basis.

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Basic and diluted EPS attributable to common shareholders was calculated as follows:

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2022	2021	2022	2021
Numerator:				
Net income (loss) attributable to common shareholders	$ 224,093	$ (30,544)	$ 178,902	$ (121,591)
Denominator:				
Weighted-average basic shares outstanding	69,445,071	68,662,673	69,305,755	68,444,061
Effect of dilutive securities				
Options to purchase common shares	937,492	—	906,866	—
Unvested RSUs	169,455	—	73,823	—
Weighted-average diluted shares	70,552,018	68,662,673	70,286,444	68,444,061
Basic EPS	$ 3.23	$ (0.44)	$ 2.58	$ (1.78)
Diluted EPS	$ 3.18	$ (0.44)	$ 2.55	$ (1.78)

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

Diluted earnings per share includes the assumed exercise of dilutive options and the assumed issuance of unvested RSUs and performance-based awards for which the performance condition has been met as of the date of determination, using the treasury stock method unless the effect is anti-dilutive. The treasury stock method assumes that proceeds, including cash received from the exercise of employee stock options and the average unrecognized compensation expense for unvested share-based compensation awards, would be used to purchase the Company's common stock at the average market price during the period.

For the three and nine months ended September 30, 2021 the Company's potentially dilutive securities, which include options and unvested RSUs, have been excluded from the computation of diluted net loss per share attributable to common shareholders for the periods indicated as the effect would be to reduce the EPS attributable to common shareholders. Therefore, the weighted average number of common shares outstanding used to calculate both basic and diluted EPS 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 EPS attributable to common shareholders for the periods indicated because including them would have had an anti-dilutive effect:

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2022	2021	2022	2021
Options to purchase common shares	8,472,937	9,556,481	8,880,055	9,556,481
Unvested RSUs	837,457	801,154	1,547,383	801,154
	9,310,394	10,357,635	10,427,438	10,357,635

13. Income Taxes

The Company is an exempted company incorporated under the laws of Bermuda. Under the current laws of Bermuda, income tax is not charged or levied on an exempted company's income. As a result, the Company has not recorded any income tax benefits from losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards will be available to the Company for those losses. The Company's wholly owned U.S. subsidiaries, Kiniksa US and Primatope, are subject to federal and state income taxes in the United States. The Company's wholly owned subsidiary Kiniksa UK, and its wholly owned subsidiaries, Kiniksa Germany, Kiniksa France, and Kiniksa

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

Although Bermuda has no corporate income tax the Company's income tax rate for the three and nine months ended September 30, 2022 is due to income subject to United Kingdom taxation and United States taxation under the Kiniksa US cost plus arrangements with the Company, and U.S. federal and state research tax credits ("R&D credits"). Income tax benefit for the three and nine months ended September 30, 2022 was $177,358 and $174,717, respectively. The benefit for income taxes is primarily driven by the release of the valuation allowance on the Company's UK deferred tax assets in the third quarter.

Management regularly reassesses the valuation allowance on the Company's deferred income tax assets. Valuation allowances require an assessment of both positive and negative evidence when determining whether it is more likely than not that the Company will be able to recover its deferred tax assets. Such assessment is required on a jurisdiction-by-jurisdiction basis. The ultimate realization of deferred tax assets is dependent upon the generation of future taxable income during the periods in which those temporary differences become deductible.

In the third quarter of 2022, the Company assessed the valuation allowance on its UK deferred tax assets and considered positive evidence, including cumulative UK income in recent years, continued commercial execution of ARCALYST, and expectations regarding future probability. After assessing both the positive evidence and negative evidence, the Company determined it was more likely than not that its UK deferred tax assets would be realized in the future and released the associated valuation allowance as of September 30, 2022. This resulted in a benefit of $185,843. As of September 30, 2022, the Company maintained a full valuation allowance against its U.S. deferred tax assets. There are no material deferred tax assets in the jurisdictions outside Kiniksa US and Kiniksa UK.

The Company transferred all of its rights, title and interest in, among other things, certain contracts (including the Biogen Agreement), intellectual property rights, product filings and approvals and other information, plans and materials owned or controlled by the Company insofar as they related exclusively or primarily to vixarelimab to Kiniksa UK in July 2022 pursuant to an asset transfer agreement between the Company and Kiniksa UK for the consideration described therein. The Company did not incur any material tax liabilities in connection with the transfer and Kiniksa UK received a stepped up basis in the property.

14. Commitments and Contingencies

License Agreements

The Company entered into license agreements with various parties under which it is obligated to make contingent and non-contingent payments (see Note 11).

Manufacturing Commitments

The Company entered into a commercial supply agreement with Regeneron to provide both clinical supply and commercial product (see Note 11). The Company entered into agreements with several CMOs to provide the Company with preclinical and clinical trial materials. As of September 30, 2022, the Company had committed to minimum payments under these agreements totaling $33,019 all of which are due within one year.

Indemnification Agreements

The Company is not aware of any claims under indemnification arrangements that are expected to have a material effect on its financial position, results of operations or cash flows, and it has not accrued any liabilities related to such obligations in its consolidated financial statements as of September 30, 2022 or December 31, 2021.

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Legal Proceedings

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

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

The following discussion and analysis of our financial condition and results of operations should be read in conjunction with our unaudited consolidated financial statements and related notes included elsewhere in this Quarterly Report on Form 10-Q, or Quarterly Report, and our audited consolidated financial statements and related notes for the year ended December 31, 2021 included in our Annual Report on Form 10-K, or Annual Report. Some of the information contained in this discussion and analysis or set forth elsewhere in this Quarterly Report, including information with respect to our plans and strategy for our business, includes forward-looking statements within the meaning of Section 27A of the Securities Act of 1933, as amended, and Section 21E of the Securities Exchange Act of 1934, as amended, that involve risks and uncertainties. As a result of many factors, including those factors set forth in the risks identified in Part II-Item 1A "Risk Factors" section of this Quarterly Report and our other filings with the Securities and Exchange Commission, or SEC, our actual results could differ materially from the results, performance or achievements expressed in or implied by these forward-looking statements.

Overview

We are a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Our portfolio of assets, ARCALYST® (rilonacept), KPL-404 and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target underserved conditions, and offer the potential for differentiation. These assets are designed to modulate immunological pathways across a spectrum of diseases.

ARCALYST is an interleukin-1α and interleukin-1β cytokine trap. In 2017, we licensed ARCALYST from Regeneron, who discovered and initially developed the drug. Our exclusive license to ARCALYST from Regeneron includes worldwide rights, excluding the Middle East and North Africa, for all applications other than those in oncology and local administration to the eye or ear. We received U.S. Food and Drug Administration, or FDA, approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older in March 2021. Recurrent pericarditis is a painful inflammatory cardiovascular disease with an estimated U.S. prevalent population of approximately 40,000 patients seeking and receiving medical treatment. ARCALYST is commercially available through a distribution network of specialty pharmacies, which provide access across the United States. ARCALYST is also approved in the United States for the treatment of Cryopyrin-Associated Periodic Syndromes, or CAPS, specifically Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome in adults and children 12 years and older, and the maintenance of remission in Deficiency of Interleukin-1 Receptor Antagonist, or DIRA, in adults and children weighing 10 kg or more. We are responsible for sales and distribution of ARCALYST in all approved indications in the United States, and evenly split profits on sales with Regeneron.

KPL-404 is an investigational monoclonal antibody inhibitor of CD40-CD154 interaction. In 2019, we acquired all of the outstanding securities of Primatope Therapeutics, Inc., or Primatope, the company that owned or controlled the intellectual property related to KPL-404. In connection with our acquisition of Primatope, we acquired an exclusive world-wide license to KPL-404 from Beth Israel Deaconess Medical Center, Inc. The CD40-CD154 interaction is a key T-cell co-stimulatory signal critical for B-cell maturation, immunoglobulin class switching and Type 1 immune response. We believe disrupting the CD40-CD154 interaction is an attractive approach to address multiple autoimmune disease pathologies such as rheumatoid arthritis, or RA, Sjogren's syndrome, Graves' disease, systemic lupus erythematosus and solid organ transplant graft rejection. In May 2021, we announced positive final data from our Phase 1 clinical trial of KPL-404 in healthy volunteers, which evaluated safety and pharmacokinetics, as well as receptor occupancy and T-cell dependent antibody response. In December 2021, we initiated a Phase 2 proof-of-concept clinical trial of KPL-404 in RA, which is designed to evaluate pharmacokinetics, safety and efficacy with subcutaneous administration. We are currently enrolling the second and final cohort of the multiple ascending dose portion of the Phase 2 clinical trial of KPL-404 in RA. Following completion of this portion of the trial, the proof-of-concept portion will begin. We expect data from the trial in the first half of 2024.

Mavrilimumab is an investigational monoclonal antibody inhibitor targeting granulocyte-macrophage colony stimulating factor receptor alpha, or GM-CSFRα. In 2017, we licensed exclusive worldwide rights in all indications to mavrilimumab from MedImmune, Limited or MedImmune. We intend to pursue collaborative study agreements to evaluate the potential of mavrilimumab in rare cardiovascular diseases where the GM-CSF mechanism has been

implicated. In parallel, we may also explore the use of mavrilimumab through research collaborations in other development areas. We previously evaluated mavrilimumab in giant cell arteritis, or GCA, a chronic inflammatory disease of the medium-to-large arteries, and COVID-19-related acute respiratory distress syndrome, or ARDS.

Vixarelimab is an investigational monoclonal antibody inhibitor of signaling through oncostatin M receptor beta, or OSMRβ. We acquired worldwide rights to vixarelimab in all indications from Biogen MA Inc., or Biogen, in 2016. In August 2022, we entered into an agreement to grant Genentech, Inc. and F. Hoffmann-La Roche Ltd (collectively, "Genentech") an exclusive worldwide license to develop and commercialize vixarelimab, which closed in September 2022. Pursuant to such agreement, we have agreed to complete our in-progress Phase 2b dose-ranging clinical trial of vixarelimab for the treatment of prurigo nodularis, a chronic inflammatory skin condition.

Our future success is dependent on our ability to continue to commercialize ARCALYST and to develop, obtain regulatory approval for and successfully commercialize one or more of our current or future product candidates. Upon approval from the FDA of the commercial marketing of ARCALYST in the United States for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older in March 2021, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States and evenly split profits on ARCALYST sales with Regeneron. However, as a company we have limited experience obtaining marketing approval for product candidates, commercializing a therapeutic, supporting sales, marketing, and distribution activities and maintaining applicable infrastructure for these activities either directly and/or through agreements with third parties; as a result we may not be able to continue to commercialize ARCALYST or successfully commercialize any future approved product candidates, if any, thus potentially impairing the commercial potential of ARCALYST and our other product candidates.

We have incurred and expect to incur significant operating losses in the future. Our ability to generate product revenue sufficient to achieve sustained corporate profitability will depend heavily on the continued commercialization of ARCALYST and the development and eventual commercialization of one or more of our current or future product candidates, if approved. While our ARCALYST collaboration with Regeneron has achieved profitability, such profits remain small compared to our total net losses and there is no guarantee that our ARCALYST collaboration with Regeneron will remain profitable in the future. In addition, payments and royalties arising from out-licensing, collaboration or other similar agreements, though potentially substantial, are often isolated events and cannot be relied upon to generate significant and sustained revenue. For the nine months ended September 30, 2022, we recognized net income of $178.9 million, primarily as a result of out-licensing activities and the release of our deferred tax asset valuation; however, as of September 30, 2022 we had an accumulated deficit of $496.5 million. Notwithstanding the foregoing, we expect to incur significant operating losses as we advance our product candidates through preclinical and clinical development and, ultimately, seek regulatory approval. In addition, we expect to continue to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution of ARCALYST. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates.

As a result, until such time as we can generate significant and sustained revenue, if ever from product sales of ARCALYST and one or more of our current or future product candidates, if approved, we expect to finance our operations through a combination of sales of ARCALYST, raising additional capital such as through debt or equity offerings or through other sources, which may include licensing, collaborations or other strategic transactions or arrangements. We may be unable to raise additional funds or enter into such other transactions or arrangements when needed on favorable terms, or at all. If we fail to raise capital or enter into such transactions or arrangements as and when needed, we may have to significantly delay, scale back or discontinue the development of one or more of our current or future product candidates, delay our pursuit of potential in-licenses or acquisitions or scale back on commercialization activities for ARCALYST.

Because of the numerous risks and uncertainties associated with product development, including any impact from the COVID-19 pandemic or fluctuations in the global economy, we are unable to predict the timing or amount of increased expenses or when or if we will be able to achieve or maintain corporate profitability. Even if we are able to continue to commercialize ARCALYST and generate product sales from one or more of our current or future product candidates, if approved, we may not become profitable or maintain profitability. If we fail to become profitable or are unable to sustain corporate profitability on a continuing basis, then we may be unable to continue our operations at planned levels and be forced to reduce or terminate our operations.

As of September 30, 2022, we had cash, cash equivalents and short-term investments of $200.7 million. We believe that our existing cash, cash equivalents and short-term investments will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months from the date of issuance of the unaudited consolidated financial statements included in this Quarterly Report. We have based this estimate on assumptions that may prove to be wrong, and we could exhaust our available capital resources sooner than we expect. See "*— Liquidity and Capital Resources*." Our future viability is dependent on our ability to fund our operations through sales of ARCALYST and/or raise additional capital, such as through debt or equity offerings, as needed.

Components of Our Results of Operations

Product revenue, net

Following the FDA approval of ARCALYST in March 2021 for recurrent pericarditis, we began generating product revenue from sales of ARCALYST in April 2021. ARCALYST is sold through a third party logistics provider that distributes primarily through a network of authorized specialty pharmacies and specialty distributors, collectively, the customers, which deliver the medication to patients by mail.

Net revenue from product sales is recognized at the transaction price when the customers obtain control of our product, which occurs at a point in time, typically upon shipment of the product from the third party logistics provider.

Our net revenues represent total revenues adjusted for discounts and allowances, including estimated cash discounts, chargebacks, rebates, returns, copay assistance, and specialty pharmacy and distributor fees. These adjustments represent variable consideration under ASC Topic 606, Revenue from Contracts with Customers, and are estimated using the expected value method and are recorded when revenue is recognized on the sale of the product. These adjustments are established by management as its best estimate based on available information and will be adjusted to reflect known changes in the factors that impact such allowances. Adjustments for variable consideration are determined based on the contractual terms with customers, historical trends, communications with customers and the levels of inventory remaining in the distribution channel, as well as expectations about the market for the product and anticipated introduction of competitive products.

License and collaboration revenue

License and collaboration revenue includes amounts recognized related to upfront payments, royalty revenue, and milestone payments.

On February 21, 2022, we entered into two collaboration and license agreements or the Collaboration Agreements, with Hangzhou Zhongmei Huadong Pharmaceutical Co., Ltd. ("Huadong"), pursuant to which we granted Huadong exclusive rights to develop and commercialize rilonacept and mavrilimumab, referred to as the Licensed Products, in the Asia Pacific region excluding Japan, or the Territory. We otherwise retained our current rights to the Licensed Products outside the Territory.

Under the Collaboration Agreements, we received a total upfront cash payment of $22.0 million, which includes $12.0 million for the Territory license of rilonacept and $10.0 million for the Territory license of mavrilimumab. In addition, we will be eligible to receive contingent payments, including specified development, regulatory and sales-based milestones. Huadong will also be obligated to pay us tiered percentage royalties on a Licensed Product-by-Licensed Product basis ranging from the low-teens to low-twenties on annual net sales of each Licensed Product in the Territory, subject to certain reductions tied to rilonacept manufacturing costs and certain other customary reductions, with an aggregate minimum floor. Royalties will be payable on a Licensed Product-by-Licensed Product and country-by-country or region-by-region basis until the later of (i) 12 years after the first commercial sale of the applicable Licensed Product in such country or region in the Territory, (ii) the date of expiration of the last valid patent claim of our patent rights or any joint collaboration patent rights that covers the applicable Licensed Product in such country or region in the Territory, and (iii) the expiration of the last regulatory exclusivity for the applicable Licensed Product in such country or region in the Territory. We recognized the $10.0 million related to the mavrilimumab license during the nine months ended September 30, 2022. We deferred the $12.0 million related to the

rilonacept license agreement as of September 30, 2022, as no materials were shipped during the nine months ended September 30, 2022.

On August 2, 2022, we entered into a license agreement (the "Genentech License Agreement") with Genentech, pursuant to which we granted Genentech exclusive worldwide rights to develop and commercialize vixarelimab and related antibodies.

Under the Genentech License Agreement, we received an upfront payment of $80.0 million for the license and are eligible to receive a near-term cash payment of $20.0 million for delivery of certain materials to Genentech. We will be eligible to receive up to approximately $600.0 million in contingent payments, including specified development, regulatory and sales-based milestones as well as royalties in the low double digits to mid-teens on annual net sales, in each case before fulfilling our upstream financial obligations. We recognized a portion of the $80.0 million upfront payment in the three and nine months ended September 30, 2022 for the exclusive vixarelimab license and the completed portion of the in-progress Phase 2b prurigo nodularis clinical trial. We will recognize the remaining revenue associated with the transaction price over the remaining duration of the in-progress Phase 2b prurigo nodularis clinical trial and after deliveries of certain drug supply.

Operating Expenses

Cost of Goods Sold

Cost of goods sold includes production and distribution costs of ARCALYST, and amortization of the $20.0 million payment we made to Regeneron in the first quarter of 2021 upon achievement of a regulatory milestone, and other miscellaneous product costs associated with ARCALYST. Cost of goods sold also includes labor and overhead costs associated with the production of ARCALYST associated with quality control, quality assurance, and supply chain activities.

Collaboration expenses

Collaboration expenses consists of Regeneron's share of the profit related to ARCALYST sales under the license agreement with Regeneron, or the Regeneron Agreement. We evenly split profits on sales of ARCALYST with Regeneron, where profits are determined after deducting from net sales of ARCALYST certain costs related to the manufacturing and commercialization of ARCALYST. Such costs include but are not limited to (i) our cost of goods sold for product used, sold or otherwise distributed for patient use by us; (ii) customary commercialization expenses, including the cost of our field force, and (iii) our cost to market, advertise and otherwise promote ARCALYST, with such costs identified in subsection (iii) subject to specified limits. In addition, should there be a transfer of technology related to the manufacture of ARCALYST, then, to the extent permitted in accordance with the Regeneron Agreement, the fully-burdened costs of each of us and Regeneron incurred in performing (or having performed) such technology transfer shall also be deducted from net sales of ARCALYST to determine profit. We also evenly split with Regeneron any proceeds received by us from any licensees, sublicensees and distributors in consideration for the sale, license or other disposition of rights with respect to ARCALYST, including upfront payments, milestone payments and royalties.

Research and Development Expenses

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

- expenses incurred to conduct the necessary preclinical studies and clinical trials required to obtain regulatory approval;

- expenses incurred under agreements with contract research organizations, or CROs that are primarily engaged in the oversight and conduct of our clinical trials and contract manufacturing organizations, or

CMOs that are primarily engaged to provide preclinical and clinical drug substance and product for our research and development programs for our product candidates;

- other costs related to acquiring and manufacturing preclinical and clinical trial materials, including manufacturing validation batches, as well as investigative sites and consultants that conduct our clinical trials, preclinical studies and other scientific development services;
- payments made in cash or equity securities under third-party licensing, acquisition and other similar agreements;
- employee-related expenses, including salaries and benefits, travel and share-based compensation expense for employees engaged in research and development functions;
- costs related to compliance with regulatory requirements; and
- allocated facilities-related costs, which include rent and utilities, depreciation and other expenses.

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

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

Research and development activities are central to our business. Product candidates in later stages of clinical development generally have higher costs than those in earlier stages of clinical development, primarily due to the increased size and duration of later-stage clinical trials. As a result, we expect that our research and development expenses will be substantial over the next several years as we conduct our ongoing and/or planned clinical trials for our product candidates as well as conduct other preclinical and clinical development, and make regulatory filings for our product candidates. We also expect to incur additional expenses related to milestone and royalty payments payable to third parties with whom we have entered into license, acquisition and other similar agreements to acquire the rights to our product candidates.

At this time, we cannot reasonably estimate or know the nature, timing and costs of the efforts that will be necessary to complete the clinical development of our current or future product candidates or when, if ever, we will realize significant revenue from product sales or be profitable. This uncertainty is due to the numerous risks and uncertainties, including those described in Part II, Item 1A. “Risk Factors” in this Quarterly Report.

Selling, General and Administrative Expenses

Selling, general and administrative expenses consist primarily of salaries and benefits, travel and share based compensation expense for personnel in selling, marketing, medical, executive, business development, finance, human resources, legal and support personnel functions. Selling, general and administrative expenses also include insurance and professional fees for legal, patent, consulting, accounting and audit services.

Upon approval from the FDA of the commercial marketing of ARCALYST in the United States for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older in March 2021, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States. We expect that our selling, general and administrative expenses will continue to increase in the future as we continue to perform commercialization and sales activities. We also anticipate that we will continue to incur significant costs, including accounting, audit, legal, compliance and director and officer insurance costs as well as investor and public relations expenses, and that such costs will increase over time as the company continues to expand.

Interest Income

Interest income consists of income recognized from investments in money market funds and U.S. Treasury notes offset by expenses related to investments.

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, there is no corporate income tax levied on an exempted company's income, resulting in an effective zero percent tax rate. As a result, we have not recorded any income tax benefits from our losses incurred in Bermuda during each reporting period, and no net operating loss carryforwards are currently available to us for those losses, while our assets remain in Bermuda. Our wholly owned U.S. subsidiaries, Kiniksa US, and Primatope are subject to federal and state income taxes in the United States. Our wholly owned subsidiary Kiniksa UK, and its wholly owned subsidiaries, Kiniksa Pharmaceuticals (Germany) GmbH, Kiniksa Pharmaceuticals (France) SARL, and Kiniksa Pharmaceuticals GmbH are subject to taxation in their respective countries. Our income tax benefit relates mainly to the release of the valuation allowance on our UK deferred tax assets partially offset by provision for income taxes relating to U.S. and UK taxable income.

In the first quarter of 2022, we transferred exclusive rights to develop and commercialize mavrilimumab in the Asia Pacific region, excluding Japan, to Kiniksa UK.

In the third quarter of 2022, we transferred exclusive worldwide rights to develop and commercialize vixarelimab to Kiniksa UK.

Results of Operations

Comparison of the Three Months Ended September 30, 2022 and 2021

The following table summarizes our results of operations for the three months ended September 30, 2022 and 2021:

	Three Months Ended September 30,		Change
	2022	2021	
		(in thousands)	
Revenue:			
Product revenue, net	$ 33,424	$ 12,095	$ 21,329
License and collaboration revenue	65,711	—	65,711
Total revenue	99,135	12,095	87,040
Costs and Operating expenses:			
Cost of goods sold	6,937	2,767	4,170
Collaboration expenses	4,623	—	4,623
Research and development	16,485	19,236	(2,751)
Selling, general and administrative	24,677	20,759	3,918
Total operating expenses	52,722	42,762	9,960
Income (loss) from operations	46,413	(30,667)	77,080
Interest income	322	5	317
Income (loss) before income taxes	46,735	(30,662)	77,397
Benefit for income taxes	177,358	118	177,240
Net income (loss)	$ 224,093	$ (30,544)	$ 254,637

Product Revenue, Net

We recognized net revenue from the sale of ARCALYST of $33.4 million for the three months ended September 30, 2022, compared to $12.1 million for the three months ended September 30, 2021, an increase of $21.3 million. The increase in product revenue was primarily driven by an increase in patients.

License and Collaboration Revenue

We reported $66.5 million of license collaboration revenue for the three months ended September 30, 2022 related to the Genentech License Agreement. We expect to recognize $14.3 million of deferred revenue related to the Genentech License Agreement over the life of the in-progress Phase 2b prurigo nodularis clinical trial, and material deliveries.

Cost of Goods Sold

Upon the first sale commencing in April 2021, we began generating cost of goods sold associated with the sales of ARCALYST. We recognized cost of goods sold from the sale of ARCALYST of $6.9 million for the three months ended September 30, 2022, compared to $2.8 million for the three months ended September 30, 2021, an increase of $4.2 million. The cost of goods sold for the three month ended September 30, 2022 and 2021 each include $0.3 million for the amortization of the payment we made to Regeneron in the first quarter of 2021 upon the achievement of a regulatory milestone. The increase in cost of goods sold relates primarily to the increase in sales and an increase in the average cost per unit. The increase in the average cost per unit is largely attributable to selling through repurposed clinical supply that was previously expensed through R&D and carried at zero-cost in the 2021 period.

Collaboration Expenses

Collaboration expenses were $4.6 million for the three months ended September 30, 2022. Our collaboration with Regeneron continued to be profitable for the three months ended September 30, 2022 after first achieving profitability in the fourth quarter of 2021, following three quarters of commercial availability of ARCALYST for recurrent pericarditis. We expect to continue to incur collaboration expenses associated with sales of ARCALYST.

Research and Development Expenses

	Three Months Ended September 30,		
	2022	2021	Change
	(in thousands)		
Rilonacept	$ 743	$ 2,178	$ (1,435)
KPL-404	3,025	1,553	1,472
Mavrilimumab	1,118	3,832	(2,714)
Vixarelimab	4,014	2,846	1,168
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	5,097	5,721	(624)
Other	2,488	3,106	(618)
Total research and development expenses	$ 16,485	$ 19,236	$ (2,751)

Research and development expenses were $16.5 million for the three months ended September 30, 2022, compared to $19.2 million for the three months ended September 30, 2021, a decrease of $2.8 million.

The direct costs for our ARCALYST program were $0.7 million during the three months ended September 30, 2022, compared to $2.2 million during the three months ended September 30, 2021, a decrease of $1.4 million. The expense reduction for the three months ended September 30, 2022 is due the limited remaining close-out costs. The expenses for the three months ended September 30, 2021 related primarily to the long-term extension portion of the RHAPSODY trial, our global, pivotal Phase 3 clinical trial in recurrent pericarditis.

The direct costs for our KPL-404 program were $3.0 million during the three months ended September 30, 2022, compared to $1.6 million during the three months ended September 30, 2021, an increase of $1.5 million. The increase in expenses incurred primarily related to the continuation costs of our Phase 2 trial in RA during the three months ended September 30, 2022 as compared to startup cost related to the Phase 2 trial in RA during the three months ended September 30, 2021.

The direct costs for our mavrilimumab program were $1.1 million during the three months ended September 30, 2022, compared to $3.8 million during the three months ended September 30, 2021, or a decrease of $2.7 million. The decrease in expenses incurred related primarily to the wind-down activities of the Phase 3 clinical trial during the three months ended September 30, 2022, while during the three months ended September 30, 2021 expenses incurred related to our Phase 2/3 clinical trial in COVID-19 related ARDS.

The direct costs for our vixarelimab program were $4.0 million during the three months ended September 30, 2022, compared to $2.8 million during the three months ended September 30, 2021, an increase of $1.2 million. Expenses incurred during the three months ended September 30, 2022 were primarily related to our continuation of the Phase 2b clinical trial in prurigo nodularis while during the three months ended September 30, 2021 expenses were primarily related to the initiation of our Phase 2b clinical trial in prurigo nodularis.

Unallocated research and development expenses were $7.6 million for the three months ended September 30, 2022 compared to $8.8 million for the three months ended September 30, 2021, a decrease of $1.2 million. The decrease of $1.2 million in unallocated research and development expenses was primarily due to a reduction

in resources to support our ongoing clinical trials. Personnel-related costs for the three months ended September 30, 2022 and 2021 included share-based compensation of $1.5 million and $1.9 million, respectively.

Selling, General and Administrative Expenses

Selling, general and administrative expenses were $24.7 million for the three months ended September 30, 2022 compared to $20.8 million for the three months ended September 30, 2021. The increase of $3.9 million was primarily due to an increase of $3.0 million in sales and marketing associated with the commercial operations of ARCALYST. Personnel-related costs for the three months ended September 30, 2022 and 2021 included share-based compensation of $4.3 million and $4.2 million, respectively.

Provision for Income Taxes

For the three months ended September 30, 2022, we recorded an income tax benefit of $177.4 million relating primarily to the release of the valuation allowance on our UK deferred tax assets. For the three months ended September 30, 2021, we recorded an income tax benefit of $0.1 million relating primarily to the tax benefit from the exercise of share options offset by the tax impact from the current tax expense due to income from our cost plus arrangements in the United States, net of R&D credits utilized. As a result of our successful commercial execution of ARCALYST we expect that our reported income tax expense will increase in future periods.

Comparison of the Nine Months Ended September 30, 2022 and 2021

	Nine Months Ended September 30,		
	2022	2021	Change
		(in thousands)	
Revenue:			
Product revenue, net	$ 82,585	$ 19,799	$ 62,786
License and collaboration revenue	75,711	—	75,711
Total revenue	158,296	19,799	138,497
Operating expenses:			
Cost of goods sold	16,185	5,233	10,952
Collaboration expenses	16,549	—	16,549
Research and development	51,100	71,864	(20,764)
Selling, general and administrative	70,736	63,207	7,529
Total operating expenses	154,570	140,304	14,266
Income (loss) from operations	3,726	(120,505)	124,231
Interest income	459	20	439
Income (loss) before benefit (provision) for income taxes	4,185	(120,485)	124,670
Benefit (provision) for income taxes	174,717	(1,106)	175,823
Net income (loss)	$ 178,902	$ (121,591)	$ 300,493

Product Revenue, Net

Following the FDA approval of ARCALYST in March 2021 for recurrent pericarditis, we began generating product revenue from sales of ARCALYST in April 2021. We recognized net revenue from the sale of ARCALYST of $82.6 million for the nine months ended September 30, 2022, compared to $19.8 million, for the nine months ended September 30, 2021, an increase of $62.8 million. The increase in product revenue was primarily driven by an increase in patients.

License and Collaboration Revenue

License and collaboration revenue for the nine months ended September 30, 2022 was $75.7 million. The license and collaboration revenue for the nine months ended September 30, 2022 primarily consisted of $10.0 million in

revenue recognized upon the signing of the mavrilimumab Collaboration Agreement in February of 2022 and $66.5 million for revenue related to the Genentech License Agreement. We expect to recognize $12.0 million of deferred revenue related to the rilonacept Collaboration Agreement over the anticipated supply period, and $14.3 million of deferred revenue related to the Genentech License Agreement over the life of the in-progress Phase 2b prurigo nodularis clinical trial, and material deliveries.

Cost of Goods Sold

Upon the first sale commencing in April 2021, we began generating cost of goods sold associated with the sales of ARCALYST. We recognized cost of goods sold from the sale of ARCALYST of $16.2 million for the nine months ended September 30, 2022, compared to $5.2 million for the nine months ended September 30, an increase of $11.0 million. The cost of goods sold for the nine months ended September 30, 2022 and 2021 each include $0.8 and $0.5 million for the amortization of the payment we made to Regeneron in the first quarter of 2021 upon the achievement of a regulatory milestone, respectively. The increase in cost of goods sold relates primarily to the increase in sales and an increase in the average cost per unit. The increase in the average cost per unit is largely attributable to selling through repurposed clinical supply that was previously expensed through R&D and carried at zero-cost during 2021.

Collaboration Expenses

Collaboration expenses were $16.5 million for the nine months ended September 30, 2022. Our collaboration with Regeneron continued to be profitable for the nine months ended September 30, 2022 after first achieving profitability in the fourth quarter of 2021, following three quarters of commercial availability of ARCALYST for recurrent pericarditis. The collaboration expenses for the nine months ended September 30, 2022 included $6.0 million due to Regeneron related to the upfront payment from the Huadong rilonacept Collaboration Agreement. We expect to continue to incur collaboration expenses associated with sales of ARCALYST.

Research and Development Expenses

	Nine Months Ended September 30,		
	2022	2021	Change
	(in thousands)		
Rilonacept	$ 1,985	$ 8,191	$ (6,206)
KPL-404	6,943	3,873	3,070
Mavrilimumab	5,607	19,749	(14,142)
Vixarelimab	10,454	7,632	2,822
Unallocated research and development expenses:			—
Personnel related (including share-based compensation)	17,276	21,804	(4,528)
Other	8,835	10,615	(1,780)
Total research and development expenses	$ 51,100	$ 71,864	$ (20,764)

Research and development expenses were $51.1 million for the nine months ended September 30, 2022, compared to $71.9 million for the nine months ended September 30, 2021, a decrease of $20.8 million.

The direct costs for our ARCALYST program were $2.0 million during the nine months ended September 30, 2022, compared to $8.2 million during the nine months ended September 30, 2021, a decrease of $6.2 million. The decrease in expense for the nine months ended September 30, 2022 is due to limited current period remaining costs offset by lower than expected RHAPSODY trial cost, our global, pivotal Phase 3 clinical trial in recurrent pericarditis, identified during the close-out activities. The expenses for the nine months ended September 30, 2021 related primarily to the long-term extension portion of the RHAPSODY trial.

The direct costs for our KPL-404 program were $6.9 million during the nine months ended September 30, 2022, compared to $3.9 million during the nine months ended September 30, 2021, an increase of $3.1 million. The increase in expenses incurred primarily related to the continuation costs of our Phase 2 trial in RA during

the nine months ended September 30, 2022, while during the nine months ended September 30, 2021 expenses incurred related primarily to the manufacturing of drug product supply for our anticipated Phase 2 trial in RA.

The direct costs for our mavrilimumab program were $5.6 million during the nine months ended September 30, 2022, compared to $19.7 million during the nine months ended September 30, 2021, a decrease of $14.1 million. The decrease in expenses incurred related primarily to the wind-down activities of the Phase 3 clinical trial in COVID-19 related ARDS during the nine months ended September 30, 2022, while during the nine months ended September 30, 2021 expenses incurred related primarily to our Phase 2/3 clinical trial in COVID-19 related ARDS.

The direct costs for our vixarelimab program were $10.5 million during the nine months ended September 30, 2022, compared to $7.6 million during the nine months ended September 30, 2021, or an increase of $2.8 million. Expenses incurred during the nine months ended September 30, 2022 were primarily related to the continuation of our Phase 2b clinical trial in prurigo nodularis while during the nine months ended September 30, 2021 expenses were primarily related to the initiation of our Phase 2b clinical trial in prurigo nodularis.

Unallocated research and development expenses were $26.1 million for the nine months ended September 30, 2022 compared to $32.4 million for the nine months ended September 30, 2021, a decrease of $6.3 million. The decrease of $6.3 million in unallocated research and development expenses was primarily due to supply chain and quality related costs associated with our commercial ARCALYST program, which are now included as cost of goods sold. Personnel-related costs for the nine months ended September 30, 2022 and 2021 included share-based compensation of $5.1 million and $6.5 million, respectively.

Selling, General and Administrative Expenses

Selling, general and administrative expenses were $70.7 million for the nine months ended September 30, 2022 compared to $63.2 million for the nine months ended September 30, 2021. The increase of $7.5 million was primarily due to an increase of $6.7 million in sales and marketing associated with the commercial operations of ARCALYST. Personnel-related costs for the nine months ended September 30, 2022 and 2021 included share-based compensation of $13.2 million and $12.5 million, respectively.

Provision for Income Taxes

For the nine months ended September 30, 2022, we recorded an income tax benefit of $174.7 million relating primarily to the release of the valuation allowance on the majority of our UK deferred tax assets. For the nine months ended September 30, 2021, we recorded a provision for income taxes of $1.1 million relating primarily to the tax impact from the current tax expense due to income from our cost plus arrangements in the United States, net of R&D credits utilized offset by tax benefit related to the exercise of share options.

Liquidity and Capital Resources

As of September 30, 2022, our principle source of liquidity was cash, cash equivalents and short-term investments, which totaled $200.7 million. Net income (loss) was $178.9 million and ($121.0) million for the nine months ended September 30, 2022 and 2021, respectively. We expect to incur operating losses for the foreseeable future.

Under various agreements with third parties, we have agreed to make milestone payments, pay royalties, annual maintenance fees and to meet due diligence requirements, each based upon specified milestones. Under our license agreement with Regeneron, we have entered into supply agreements to provide both clinical and commercial product. We have committed to minimum payments to Regeneron of $32.8 million, all of which are due within one year. We have entered into lease agreements for office and laboratory space, and vehicles, with total future lease payments of $7.0 million, of which $3.5 million are due within one year.

Under various agreements with third parties, we are entitled to receive upfront payments, milestone payments, and royalties, each based upon specified milestones. In the second quarter of 2022 we received $22.0 million of upfront

payments as part of the Collaboration Agreements with Huadong. In the third quarter of 2022 we received an $80 million upfront payment as part of the Genentech License Agreement.

These agreements impact our short-term and long-term liquidity and capital needs.

Cash Flows

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

	Nine Months Ended September 30,	
	2022	2021
	(in thousands)	
Net cash provided by (used in) operating activities	$ 17,383	$ (107,414)
Net cash provided by investing activities	34,582	178,676
Net cash provided by financing activities	1,326	4,673
Net increase (decrease) in cash and cash equivalents and restricted cash	$ 53,291	$ 75,935

Operating Activities

During the nine months ended September 30, 2022, operating activities provided $17.4 million of cash, primarily resulting from our net income of $178.9 million excluding the impact of deferred income taxes and other noncash charges of $162.9 million. Noncash charges and the benefit from deferred income taxes for the nine month ended September 30, 2022 consisted primarily of an increase to deferred income tax assets of $185.8 million offset by $18.7 million of stock based compensation. Net cash provided by our operating assets and liabilities for the nine months ended September 30, 2022 consisted primarily of a change in deferred revenue of $26.3 million as a result of the rilonacept Huadong Collaboration Agreement and the Genentech License Agreement, offset by a $10.9 million increase in inventory, a $8.7 million increase in prepaid expenses and other current assets, and a $7.2 million increase in accounts receivable.

During the nine months ended September 30, 2021, operating activities used $107.4 million of cash, primarily resulting from our net loss of $121.6 million as well as net cash used by our operating assets and liabilities of $9.0 million offset by non-cash charges of $23.2 million. Net cash used by our operating assets and liabilities for the nine months ended September 30, 2021 consisted primarily of a $5.6 million increase in inventory and a $3.2 million increase in accounts receivable primarily due to the commercial launch of ARCALYST in 2021 offset by an increase of $1.4 million in accrued expenses and accounts payable primarily due to increases related to our clinical trial costs and other selling, general and administration accruals offset by our pre-commercialization activities of our ARCALYST program and the cash payment of the 2020 employee bonuses.

Investing Activities

During the nine months ended September 30, 2022 investing activities provided $34.6 million of cash, primarily consisting of $107.7 million from proceeds of maturities of short-term investments, partially offset by $73.1 million of purchases of short-term investments.

During the nine months ended September 30, 2021 investing activities provided $178.7 million of cash, consisting of $296.3 million from proceeds of maturities of short-term investments, partially offset by $97.5 million of purchases of short-term investments and $20.0 million related to the payment of a regulatory milestone incurred under the Regeneron Agreement.

Financing Activities

During the nine months ended September 30, 2022 and 2021, net cash provided by financing activities was $1.3 million and $4.7 million, respectively, consisting of proceeds from the exercise of share options.

Funding Requirements

We expect to incur significant expenses in connection with our ongoing and planned activities as we continue to commercialize ARCALYST and advance our current and future product candidates through preclinical and clinical development, seek regulatory approval and commercialize one or more of our current or future product candidates, if approved. In addition, if we obtain marketing approval for any of our current or future product candidates, we expect to incur significant additional commercialization expenses related to such activities. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates. As a result, we expect to incur additional expenses related to milestone, royalty and other payments payable to third parties with whom we have entered into license, acquisition and other similar agreements to acquire the rights to our product candidates. Additionally, we expect to continue to incur costs associated with operating as a public company, including significant legal, accounting, investor relations and other expenses. We expect to incur expenses as we:

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

We believe that our existing cash, cash equivalents and short-term investments will enable us to fund our operating expenses and capital expenditure requirements for at least the next 12 months. The future viability of our company is dependent on our ability to fund our operations through sales of ARCALYST and/or raise additional capital, such as through debt or equity offerings, as needed. We anticipate that we may require additional capital if we choose to pursue in-licenses or acquisitions of other product candidates and technologies or their related businesses. We expect to continue to incur significant expenses related to product manufacturing, sales, marketing and distribution of ARCALYST. In addition, if we obtain regulatory approval for any of our current or future product candidates, pursue additional indications or additional territories for our products or any of our current or future product candidates, we expect to incur significant expenses related to product development and manufacturing, sales, marketing and distribution, depending on where we choose to commercialize.

Because of the numerous risks and uncertainties associated with research, development and commercialization of biologic products, we are unable to estimate the exact amount of our working capital requirements. Our future funding requirements may be impacted by a number of factors, including those described in Part II, Item 1A. “Risk Factors” in this Quarterly Report.

Until such time, if ever, as we can generate substantial and sustained product revenue, we expect to finance our cash needs through a combination of public or private equity offerings, debt financings, or other sources, including, licensing, collaboration, marketing, distribution or other strategic transactions or arrangements with third parties. To the extent that we raise additional capital through the sale of equity or convertible debt securities, our shareholders’ ownership interest may be materially diluted, and the terms of such securities could include liquidation or other preferences that adversely affect our shareholders’ rights as a common shareholder. Debt financing and preferred equity financing, if available, may involve agreements that include restrictive covenants that limit our ability to take specified actions, such as incurring additional debt, making capital expenditures or declaring dividends. In addition, debt financing would result in fixed payment obligations.

If we raise funds through licensing, collaboration, marketing, distribution or other strategic transactions or arrangements with third parties, we may have to relinquish valuable rights to our technologies, product candidates or future revenue streams, or otherwise agree to terms that may not be favorable to us. If we are unable to obtain funding, we could be forced to delay, reduce or eliminate some or all of our research and development programs for product candidates, product portfolio expansion or commercialization efforts, which could adversely affect our business prospects, or we may be unable to continue operations.

Critical Accounting Policies and Significant Judgments and Estimates

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

During the nine months ended September 30, 2022, we included the estimates associated with revenue recognition in our critical accounting policies. Our critical accounting policies are described under the heading “Management’s Discussion and Analysis of Financial Condition and Results of Operations—Critical Accounting Policies and Significant Judgments and Estimates” in the Annual Report and the notes to the consolidated financial statements included in Item 1, “Consolidated Unaudited Financial Statements,” included in this Quarterly Report. We believe that of our critical accounting policies, the following accounting policies involve the most judgment and complexity:

- accrued research and development expenses;
- share-based compensation;
- revenue recognition; and
- realizability of deferred tax assets.

Item 3. Quantitative and Qualitative Disclosures About Market Risk.

Interest Rate Risk

We are exposed to market risk related to changes in interest rates. As of September 30, 2022, our cash, cash equivalents and short-term investments consisted of money market funds and U.S. Treasury notes. Our primary exposure to market risk is interest income sensitivity, which is affected by changes in the general level of U.S. interest rates. Further, the COVID-19 pandemic and the ongoing war in Ukraine have adversely impacted the U.S. and global economy and financial markets, and any prolonged impact may continue to affect market interest rates. However, because of the short-term nature of the instruments in our portfolio, an immediate 10% change in market interest rates would not have a material impact on the fair market value of our investment portfolio or on our financial position or results of operations.

Item 4. Controls and Procedures.

Limitations on Effectiveness of Controls and Procedures

In designing and evaluating our disclosure controls and procedures, management recognizes that any controls and procedures, no matter how well designed and operated, can provide only reasonable assurance of achieving the desired control objectives.

Evaluation of Disclosure Controls and Procedures

Our management, with the participation of our principal executive officer and principal financial officer, evaluated, as of the end of the period covered by this Quarterly Report, the effectiveness of our disclosure controls and procedures (as defined in Rules 13a-15(e) and 15d-15(e) under the Securities Exchange Act of 1934, as amended, or the Exchange Act). Based on that evaluation, our principal executive officer and principal financial officer concluded that our disclosure controls and procedures were effective at the reasonable assurance level as of September 30, 2022.

Changes in Internal Control over Financial Reporting

There was no change in our internal control over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act) during the three months ended September 30, 2022 that has materially affected, or is reasonably likely to materially affect, our internal control over financial reporting.

PART II - OTHER INFORMATION

Item 1. Legal Proceedings.

We are not party to any material legal proceedings.

Item 1A. Risk Factors.

You should carefully consider the risks described below, as well as the other information in this Quarterly Report, including our unaudited consolidated financial statements and the related notes and "Management's Discussion and Analysis of Financial Condition and Results of Operations." The occurrence of any of the events or developments described below could adversely affect our business, financial condition, results of operations and growth prospects. In such an event, the market price of our Class A common shares could decline. Additional risks and uncertainties not presently known to us or that we currently deem immaterial also may impair our business operations.

Risks Related to Our Financial Position and Capital Needs

We are a biopharmaceutical company and have only started to generate revenue from product sales. We have incurred significant operating losses since our inception and anticipate that we will incur continued losses for the foreseeable future.

Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk. Typically, it takes many years to develop one new product from the time it is discovered to when it is available for treating patients and development may cease for a number of reasons. We have incurred corporate operating losses in each year since our inception in 2015 and anticipate incurring losses for the foreseeable future. Our future success is dependent on our ability to develop, obtain regulatory approval for and successfully commercialize one or more of our product candidates. While the FDA approved ARCALYST® (rilonacept) for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021, we may not be able to continue to commercialize ARCALYST or obtain significant and sustained revenue from such commercialization.

To date, we have incurred significant losses related to expenses for research and development and our ongoing operations. For the nine months ended September 30, 2022, we recognized net income of $178.9 million as a result of out-licensing activities; however, as of September 30, 2022, we had an accumulated deficit of $496.5 million. We expect to incur losses for the foreseeable future as a result of many factors, including:

- supporting our sales, marketing and distribution capabilities, infrastructure and organization to commercialize ARCALYST and any products candidates for which we may obtain marketing approval for indications in the United States;
- our research and preclinical and clinical development of our product candidates, including our Phase 2b dose-ranging study of vixarelimab in prurigo nodularis and our Phase 2 proof-of-concept clinical trial for KPL-404 in RA;
- manufacturing our products and product candidates for clinical or commercial use, and increasing our manufacturing capabilities or adding additional manufacturers or suppliers;
- seeking regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;
- initiating potential additional preclinical studies and clinical trials for our product candidates;
- making milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreement;

- seeking to identify, assess and study new or expanded indications for our products or product candidates, new or alternative dosing levels and frequency for our products or product candidates, or new or alternative administration of our products or product candidates, including method, mode or delivery device;
- seeking to identify, assess, acquire or develop additional product candidates;
- entering into licensing, acquisition, collaboration or other strategic transaction agreements;
- seeking to maintain, protect and expand our intellectual property portfolio;
- seeking to attract and retain skilled personnel;
- creating additional infrastructure to support our product development and commercialization efforts; and
- experiencing delays or encountering issues with any of the above, including but not limited to the impact of the COVID-19 pandemic and measures taken in response to the pandemic, failed trials, complex results, safety issues, regulatory challenges that require longer follow-up of existing trials, additional major trials, additional supportive trials in order to pursue marketing approval or the global economic slowdown and rising inflation.

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

Further, the net losses we incur may fluctuate significantly from quarter-to-quarter and year-to-year, such that a period-to-period comparison of our results of operations may not be a good indication of our future performance. Even if we achieve corporate profitability in the future, we may not be able to sustain such 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 (deficit) and working capital.

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

The development and commercialization of biopharmaceutical products is capital intensive. We are currently commercializing ARCALYST in the United States for the treatment of recurrent pericarditis, CAPS and DIRA. In addition, we are advancing our product candidates through research, preclinical and clinical development, including our Phase 2 clinical trial with KPL-404 in RA and our Phase 2b dose-ranging study of vixarelimab in prurigo nodularis.

Our expenses may increase in connection with our ongoing activities as we continue to support our sales, marketing and distribution capabilities, continue the research and development of our product candidates and expand our infrastructure and organization to support such activities. We also may incur significant additional commercialization expenses with respect to any future marketing approval of any of our product candidates related to manufacturing, product sales, marketing and distribution. As our product candidates progress through development and towards potential commercialization, we will need to make milestone payments and, if successful, eventually make royalty payments to the applicable licensors and other third parties from whom we have acquired our product candidates. Furthermore, we expect to continue to incur costs associated with operating as a public company.

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

Our business is highly uncertain, and we cannot estimate with certainty the actual amounts necessary to successfully market and sell products, or complete the development, regulatory approval process and commercialization of our product candidates. Our operating plans may change as a result of many factors currently unknown to us, and we may need to seek additional funds sooner than expected, through public or private securities offerings, debt financings or other sources, including government funding or grants. Such factors that may significantly impact our funding requirements include:

- our ability to continue to commercialize ARCALYST or successfully commercialize any of our current or future product candidates, if approved, including the cost and timing of supporting our sales, marketing and distribution capabilities, infrastructure and organization expansion and entering into agreements with third parties to conduct one or more of these activities;

- the amount and timing of sales revenues from ARCALYST or any of our product candidates, if approved in the future, including the sales price and the availability of coverage and adequate third-party reimbursement;

- competitive and potentially competitive products and technologies and patients' and prescribers' receptivity to ARCALYST or any of our product candidates if approved in the future and the technology underlying them in light of competitive products and technologies;

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

- the results from, and the time and cost necessary for development of our product candidates;

- the costs and timing of establishing and maintaining clinical trial sites for the development of our product candidates, both in the United States and in jurisdictions outside of the United States, including as a result of the COVID-19 pandemic and global political turmoil, including the ongoing war in Ukraine;

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

- the costs, timing and outcomes of seeking and potentially obtaining approvals from regulatory authorities, including the potential for regulatory authorities to require that we conduct more studies than we currently plan to conduct and the costs of conducting post-marketing studies or implementing a Risk Evaluation and Mitigation Strategy, or REMS, that could be required by regulatory authorities;

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

- the timing and amount of milestone and other payments we may receive under our agreements with Huadong, Genentech and any other third parties to whom we may in the future out-license products and product candidates;

- the costs to identify, assess and study new or expanded indications for our products and product candidates, new or alternative dosing levels or frequency for our products or product candidates, or new or alternative administration of our products or product candidates, including method, mode or delivery device;

- the costs of any future in-license, acquisition, development or discovery of additional product candidates, including in connection with any licensing, acquisition, collaboration or other strategic transaction agreements;
- the 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 products and product candidates or any related activities;
- the ongoing costs associated with being a public company;
- our need and ability to hire and retain skilled personnel; and
- the receptivity of the capital markets to financings by biopharmaceutical companies generally and companies with a single commercial product and product candidates and technologies such as ours, specifically.

Any additional fundraising efforts may divert our management from their day-to-day activities, which may adversely affect our ability to develop and commercialize our product candidates.

In addition, the COVID-19 pandemic and the ongoing war in Ukraine continues to adversely impact the global economy, posing risks to our business and the business of third-parties upon whom we rely. See "*Risk Factors — General Risk Factors — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position"* and "*Risk Factors — General Risk Factors — The ongoing war in Ukraine, and actions taken against Russia as a result of its invasion of Ukraine, has and may continue to have an adverse impact on the global economy, equity capital markets and our clinical operations."*

Additionally, funds may not be available when we need them, on terms that are acceptable to us, or at all. If we are unable to obtain funding when needed, we will be forced to curtail, delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or commercialization efforts of any of our products or product candidates for which we obtain approval. 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 or products.

If and until such time as we can generate substantial and sustained product revenue from ARCALYST or any of our product candidates, if approved in the future, we expect to finance most of our cash needs through private or public securities offerings, debt financings, government funding or grants, or other sources, including licensing, collaboration or other strategic transactions or arrangements with third parties. The terms of any financing may adversely affect the holdings or the rights of our shareholders and our issuance of additional securities, whether equity or debt, or the possibility of such issuance, may cause the market price of our Class A common shares to decline. The sale of additional equity or convertible securities would dilute all of our shareholders. The incurrence of indebtedness would result in increased fixed payment obligations and we may be required to agree to certain restrictive covenants, such as limitations on our ability to incur additional debt, limitations on our ability to acquire, sell or license intellectual property rights and

other operating restrictions that could adversely impact our ability to conduct our business. Obtaining funds through licensing, collaboration or other strategic transactions or arrangements with third parties may require us to relinquish rights to some of our technologies, product candidates or future revenue streams, or otherwise agree to terms unfavorable to us, any of which may have a material adverse effect on our business, operating results and prospects. If we raise funds through research grants, we may be subject to certain requirements, which may limit our ability to use the funds or require us to share information from our research and development. Raising additional capital through any of these or other means could adversely affect our business and the holdings or rights of our shareholders, and may cause the market price of our Class A common shares to decline.

Risks Related to Commercialization

We have limited experience as a company commercializing a therapeutic product and supporting sales, marketing, distribution and general infrastructure either directly and/or through agreements with third parties. As a result, we may not be able to continue to commercialize ARCALYST or be successful in commercializing any future approved product candidates, potentially impairing commercial potential for ARCALYST and our product candidates to generate any revenue.

We have been commercializing ARCALYST since April 2021. Since that time, we have focused on establishing and expanding our internal capabilities, including but not limited to, sales, marketing, distribution, access and patient support services as well as contracting with third parties to perform certain services. Each aspect of commercialization on its own can be complex, expensive, and time consuming, and, collectively, the required effort for coordination is intensive. While we are more than a year into the commercialization of ARCALYST and have realized revenues from such efforts, there is no guarantee that we will be able to continue our commercialization of the product or be able to maintain the trajectory of growth or significant and sustained revenues in the long-term.

In addition, our continued commercialization of ARCALYST or successful commercialization of any of our current or future product candidates, if approved, is subject to a number of foreseen and unforeseen factors, including:

- our inability to recruit, train and retain adequate numbers of effective sales, marketing, access, and payor and patient support personnel;
- the inability of sales personnel to obtain access to prescribers and accounts as well as for an adequate number of prescribers or accounts to prescribe any of our future products;
- the lack of complementary products to be supported by sales personnel, which may put us at a competitive disadvantage relative to companies with more extensive product lines;
- our inability to develop strong scientific-based relationships to drive disease awareness and education;
- our inability to establish the unmet medical need for a given disease;
- our inability to enable our products to be viewed as the product of choice within any indications for which they are approved;
- our inability or delay in gaining or maintaining reimbursement and broad patient access at a price that reflects the value of ARCALYST or any of our future products;
- our inability to equip customer-facing personnel with effective materials, including medical and sales literature to help them educate physicians and other healthcare providers regarding applicable diseases relevant to ARCALYST or any of our future products;

- any delays in our ability to produce sufficient quantities of ARCALYST, or any of our future products, at an acceptable cost or quality, including such delays arising out of quality assurance concerns or changes in regulatory guidance, or those caused by our reliance on our third-party manufacturers;

- our inability and the inability of any third parties upon which we rely to effectively distribute products in a timely manner;

- our inability to provide prescribers and patients adequate support and training to build comfort around the preparation and administration process to initiate and continue to use ARCALYST or any of our future products;

- our inability to develop robust patient support programs to optimize the patient and customer experience with ARCALYST or any of our future products;

- our inability to develop or obtain and sustain sufficient operational functions and infrastructure to support our commercial activities; and

- unforeseen costs and expenses associated with creating and maintaining a sales, marketing, and access organization.

In addition, disruptions caused by the evolving COVID-19 pandemic may increase the likelihood that we encounter such difficulties, delays or unforeseen costs. If we experience any such factors that inhibit our efforts to commercialize ARCALYST or any of our product candidates, if approved, our business, results of operations, financial condition and prospects may be materially adversely affected. See "*Risk Factors — General Risk Factors — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position.*"

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

Even with FDA or any other regulatory authority approval of the marketing of ARCALYST or any of our other product candidates in the future (whether developed on our own or with a collaborator), prescribers, healthcare providers, patients, the medical community or third-party payors may not accept or use ARCALYST or any of our future product candidates, or may effectively block or limit their use in the case of third-party payors. While ARCALYST has seen near-term success, it is not certain we will be able to sustain such success over the long-term. If ARCALYST or any of our other product candidates, if approved, do not achieve an adequate level of sustained acceptance, we may not generate a sufficient level of product revenue or profits from operations, if at all. Sustained market acceptance of ARCALYST in the approved recurrent pericarditis indication, or any of our future approved product candidates and continued use of such products by our patients, will depend on a variety of factors, including:

- the timing of market introduction;

- disease awareness, including understanding the severity and epidemiology of the disease;

- the number and clinical profile of competing products, whether approved or not;

- the potential and perceived advantages or disadvantages of our product candidates relative to alternative treatments;

- 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 regulatory authorities;
- limitations or warnings contained in the labeling approved by regulatory authorities;
- convenience and ease of administration, including relative to alternative therapies;
- pricing (including patient out-of-pocket costs), budget impact, affordability and cost effectiveness, particularly in relation to alternative treatments;
- market acceptance of current and future price increases of our products;
- the effectiveness of our sales, marketing and distribution activities;
- availability of adequate coverage, reimbursement and payment from health maintenance organizations and other insurers, both public and private, and the timing thereof; and
- other potential advantages over alternative treatment methods.

If ARCALYST or any of our future approved products, if any, fail to gain market acceptance, our ability to generate revenue will be adversely affected. Even if ARCALYST or any future approved product candidates achieve market acceptance, the relevant market may prove not to be large enough to allow us to generate significant and sustained revenue.

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

Our ability to continue to commercialize ARCALYST in the approved recurrent pericarditis indication and other approved indications in the United States or any of our future approved product candidates, if any, particularly in orphan or rare disease indications, will depend in part on the availability of favorable coverage, patient affordability and the adequacy of reimbursement for ARCALYST or the product candidate and alternative treatments from third-party payors (e.g., governmental authorities, private health insurers and other organizations). We currently enjoy favorable coverage and reimbursement from third-party payors for ARCALYST in the approved recurrent pericarditis indication and seek to maintain such favorable coverage and reimbursement.

We cannot be certain we will continue to effectively execute our coverage and reimbursement strategy in the markets we pursue, which could limit the future commercial potential of ARCALYST in the approved recurrent pericarditis indication or any of our product candidates, if approved.

Governmental authorities, private health insurers and other third-party payors have attempted to control costs through a number of efforts, including by delaying the time to reimbursement, by restricting the breadth of coverage, by limiting the amount of reimbursement for particular products in terms of lower pricing and by increasing the proportion of the cost for which the patient is responsible. There may be significant delays in obtaining reimbursement for newly approved products or product indications, coverage may be limited to a subset of the patient population for which the treatment is approved by the FDA or similar regulatory authorities outside the United States, and reimbursement rates may vary according to the use of the product and the clinical setting in which it is used. Coverage and reimbursement barriers by payors may materially impact the demand for, or the price of, ARCALYST and any product candidate for

which we obtain marketing approval, if any. If coverage and reimbursement are not available, or available only at limited levels, or if such coverage will require patient out-of-pocket costs that are unacceptably high, we may not be able to successfully commercialize ARCALYST or any of the product candidates for which we obtain marketing approval. Moreover, any coverage or reimbursement that may be obtained may be decreased or eliminated in the future. For example, one of the large private health insurers that currently covers ARCALYST intends to place ARCALYST on its exclusion list for the CAPS indication, which could create hurdles for new patients seeking coverage for their prescriptions in all indications.

We may also be unable to adequately satisfy a third-party payor's value/benefit assessment on an ongoing basis. It is possible that third-party payors will select low-cost clinical comparators that serve as benchmarks for determining relative value, including generics, biosimilars and lower costs brands with or without the same approved indication. The result of such a change would be a more challenging value/benefit assessment caused by a more challenging basis for comparison and the potential for a worse relative outcome. Third-party payors may determine that we have failed to generate sufficient evidence to demonstrate the relative benefits of ARCALYST or any of our product candidates, if approved, and refuse to provide coverage and reimbursement entirely, or many find the evidence not sufficiently compelling to support the desired pricing and reimbursement. Similarly, payors may implement coverage criteria that further restricts the use of ARCALYST or any of our product candidates, if approved, beyond the approved label, which could adversely affect their commercial potential, including, for example, situations where a patient must be proven to not adequately respond to the lower-cost comparator.

Third-party payors are also introducing more challenging price negotiation methodologies, including in re-visiting established coverage and reimbursement parameters when new competitors, including branded drugs, generics and biosimilars, enter the market. It is possible that a third-party payor may consider our products and product candidates, if approved, as substitutable and only be willing to cover the cost of the alternative product. Even if we show improved efficacy, safety or improved convenience of administration with ARCALYST or any of our product candidates, if approved, pricing of competitive products may limit the amount we will be able to charge for ARCALYST or any of our product candidates, if approved. Third-party payors may deny or revoke the reimbursement status of a given product or establish prices for new or existing marketed products at levels that are too low to enable us to realize an appropriate return on our investment in our product candidates. In some cases, when new competitor generic and biosimilar products enter the market, there are mandatory price reductions for the innovator compound. In other cases, payors employ "therapeutic category" price referencing and seek to lower the reimbursement levels for all treatment in the respective therapeutic category. Additionally, new competitor brand drugs can trigger therapeutic category reviews in the interest of modifying coverage and/or reimbursement levels. The potential of third-party payors to introduce more challenging price negotiation methodologies could have a negative impact on our ability to continue to commercialize ARCALYST or successfully commercialize any of our product candidates, if approved. Third-party payors may also employ challenging price negotiation tactics in the event of a proposed price increase of our current and future products. See "Risks Related to Commercialization – *It may be difficult for us to realize the benefit of increasing the price of certain of our commercialized products, due to unfavorable actions that third-party payors and other market participants may take.* "

It may be difficult for us to realize the benefit of increasing the price of certain of our commercialized products, due to unfavorable actions that third-party payors and other market participants may take.

In July 2022, we increased the price of ARCALYST, the first price increase of such drug since its initial commercialization for CAPS in 2008. In the future we may continue to increase the price of ARCALYST or any of our future products. We may be unable to realize commercial benefits from such price increases due to unfavorable actions that third-party payors (including governmental authorities and private health insurers) may take in response to price increases. Even if price increases lie below contractual price protection clauses, payors may request price concessions in exchange for covering our products or may opt to change coverage or reimbursement policies with respect to such products. If we cannot successfully negotiate with such payors, we may be forced to provide significant price concessions or, if we fail to arrive at a satisfactory resolution, lose favorable coverage or reimbursement for patients served by such payor. In such an event, we may see materially negative impacts on the patients we seek to serve and our business operations.

Any price concessions will reduce our overall revenue generation and may impair the benefit of any price increases we may take. As an early-commercial stage biopharmaceutical company, we have not yet achieved corporate profitability and our future success relies on the effective commercialization of one or more products. Price concessions that reduce potential product revenue would lengthen our timeline to profitability and may require us to rely on potentially dilutive capital-raising efforts to fund our operations, which may impact the price of our common shares. Even comparatively small discounts, if aggregated across payors, may cause materially lower revenue generation in the long-term which may offset the increased revenue we hoped to realize through a price increase.

Further, granting price concessions to one or more payors may limit our ability to negotiate prices with other payors or in other territories. Payors, including governmental payors, negotiate drug prices by reference to the prices we have set with other payors. Should payors become aware of price concessions that we have granted, they may request similar concessions. If enough payors request and receive price concessions, our ability to generate revenue may be materially impacted, harming our business, financial condition and results of operations. Further, this may limit our ability to secure acceptable prices in potential new territories, which may materially limit our overall commercial growth. A limitation on our ability to commercialize in new and existing territories may also potentially reduce our access to the patient populations we seek to serve, harming our ability to deliver therapeutics to patients with unmet need.

In the event that we cannot successfully negotiate with payors requesting price concessions in connection with a price increase or otherwise, such payors may choose to not cover our current and future products at all or may instate onerous reimbursement policies that limit patient access. We cannot assure you that current payor coverage and reimbursement policies for ARCALYST will continue. As an early-commercial stage company, the loss of any payor, especially a large payor, or limitations on patient access to our drugs affecting a sizeable number of patients may materially harm our ability to generate revenue and execute on our commercial strategy. Further, as a company targeting patients with significant unmet need, the loss of access to our products may materially harm our targeted patient populations who cannot source adequate alternative therapies.

Further, price increases that outpace inflation may trigger additional rebate obligations under the Medicaid drug rebate program, Medicare Part B and Medicare Part D.

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

The precise incidence and prevalence for all the conditions we aim to address with our programs are not known with specificity. Our projections of both the number of people who have these diseases, as well as the subset of people with these diseases who have the potential to benefit from treatment with our products and product candidates, if approved, are based largely on our extrapolation from available population studies and estimates. These estimates have been derived from a variety of sources, including scientific literature, surveys of clinics, patient foundations, pharmacy claims analyses, large national surveillance databases or market research, and may prove to be incorrect. Further, new trials and therapeutic options may lead to changes in the estimated incidence or prevalence of these diseases, or relevant subpopulations thereof. As a result, the number of patients who may benefit from our products or product candidates, if approved, may turn out to be lower than expected.

The total addressable market for any of our products and approved product candidates in the future, if any, will ultimately depend upon, among other things, the diagnostic criteria and applicable patient population included in the final label for the product or product candidate approved for sale for its indication, the efficacy, safety and tolerability demonstrated by the product candidate in our clinical trials, acceptance by the medical community and patients, pricing, access and reimbursement. The number of addressable patients in the United States and other major markets outside of the United States may turn out to be lower than expected, patients may not be otherwise amenable to treatment with our products or new patients may become increasingly difficult to identify or gain access to, all of which would adversely affect our results of operations and our business. Further, even if we obtain significant market share for our product

candidates, because the potential target populations are small for many of our approved and targeted indications, we may never achieve corporate profitability.

Evolving health policy and associated legislative changes related to coverage and reimbursement aimed at lowering healthcare expenditure could impact the commercialization of our product candidates. Pharmaceutical pricing has been, and likely will continue to be, a central component of these efforts.

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

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

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

As a result of the foregoing, we may not be able to achieve or sustain favorable pricing for ARCALYST or any of our product candidates, if approved, and adequate reimbursement, which may hinder our ability to recoup our investment in such drugs.

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

We face an inherent risk of product liability exposure related to the commercialization of ARCALYST and the testing of our product candidates in clinical trials and other research activities. If we cannot successfully defend ourselves against claims that our products or 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 or products we commercialize;
- injury to our reputation and significant negative media attention;
- regulatory investigations that could require costly recalls or product modifications;
- withdrawal of clinical trial participants;
- significant costs to defend the related litigation;
- substantial monetary awards to trial participants or patients;

- loss of potential revenue;
- the diversion of management's attention away from managing our business; and
- the inability to commercialize any product candidates that we may develop, if approved.

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. 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.

If, in the future, we are unable to maintain our sales, marketing and distribution capabilities, infrastructure and organization directly and/or through agreements with third parties to sell and market our products and product candidates, if approved, their commercial potential may be impaired.

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

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 approved product candidates ourselves. In addition, we may not be successful in entering into arrangements with third parties to market and sell our approved product candidates, if any, or may be unable to do so on terms that are favorable to us. Further, we will likely 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 approved product candidates effectively. However, developing a sales, marketing and access organization requires significant investment, is time consuming and if not completed as planned could delay the launch of our approved product candidates. Furthermore, we may not be able to adequately establish an effective sales, marketing, distribution and access organization in the European Union, or the EU, or other key markets in which we may obtain approval for the commercial marketing of our product candidates outside of the United States. If we are unable to maintain or establish sales, as applicable, marketing and distribution capabilities successfully, either on our own or in collaboration with third parties, we will not be successful in commercializing our approved product candidates, if any, and the approved product candidates' ability to generate any revenue may be impaired. Furthermore, our business, results of operations, financial condition and prospects will be materially adversely affected.

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

Our future corporate profitability may depend, in part, on our ability to commercialize our product candidates in markets outside of the United States for which we may rely on collaborations with third parties.

We continue to evaluate the opportunities for the development and commercialization of our product candidates in certain markets outside of the United States, including through our Managed Access Program and collaborations with third parties, including Huadong. We and our collaborators are not permitted to market or promote any of our product candidates before we receive regulatory approval from the applicable regulatory authority in that market, and we may never receive such regulatory approval for any of our product candidates. To obtain separate regulatory approval in many other countries, we, or our collaborators, must comply with numerous and varying regulatory requirements of such countries regarding safety and efficacy and governing, among other things, clinical trials, manufacturing and commercial sales, pricing and distribution of our product candidates, and we cannot predict success in these jurisdictions. If we

obtain approval of our product candidates and ultimately commercialize our product candidates in markets outside of the United States, we would be subject to additional risks and uncertainties, including:

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

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

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

We may also be subject to burdensome pricing requirements. See "*Risk Factors – Risks Related to Commercialization –Evolving health policy and associated legislative changes related to coverage and reimbursement aimed at lowering healthcare expenditure could impact the commercialization of our product candidates. Pharmaceutical pricing has been, and likely will continue to be, a central component of these efforts.*"

We are subject to ongoing obligations, regulatory requirements and continued regulatory review, which may result in significant additional expense. Additionally, our products and future approved product candidates, if any, could be subject to unfavorable changes 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.

We are subject to ongoing regulatory requirements for a number of our activities, including manufacturing, packaging, labeling, storage, distribution, advertising, promotion, sampling, record-keeping, adverse event reporting,

conduct of post-marketing trials and submission of safety, efficacy and other post-market information for our products in the United States. Such obligations, along with continued regulatory review, may result in significant additional expense. Furthermore, if we seek and receive approval from regulatory authorities outside of the United States for products or any of our product candidates in the future, we will be subject to such authorities' requirements, which may be over and above our obligations in the United States.

Manufacturers and their facilities are required to comply with extensive requirements of regulatory authorities, including ensuring that quality control and manufacturing procedures conform to current good manufacturing practices, or cGMP, or similar foreign regulations. As such, we and our contract manufacturing organizations, or CMOs, will be subject to user fees and continual review and inspections to assess compliance with cGMP or similar foreign regulations and adherence to commitments made in any biologics license applications, or BLA, or Marketing Authorization Application, or MAA. Accordingly, we and our CMOs and others with whom we work must continue to expend time, money and effort in all areas of regulatory compliance, including manufacturing, production and quality control.

Any regulatory approvals that we receive 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 safety and efficacy. For example, the holder of an approved BLA or similar foreign application 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 our marketing approval would be 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 or foreign regulatory authority also may place other conditions on approvals including the requirement for a REMS or similar risk management measures, to assure the safe use of the product. If the FDA or foreign regulatory authority concludes a REMS or similar risk management measures are needed, the sponsor of the BLA or MAA must submit a proposed REMS or the similar risk management measures 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. We also will be required to report certain adverse reactions, production problems, inadequate efficacy and other issues, if any, to applicable regulatory authorities on an ongoing basis. In addition, the identification of new safety issues could lead to new labeling or restrictions on population or use of our products, diminishing the addressable market or sales or both. Such conditions, requirements or events may prove to be expensive and burdensome, and the reporting of such may cause the price of our Class A common shares to decrease.

Further, we must also comply with additional requirements concerning advertising and promotion for our products, which are subject to a variety of legal and regulatory restrictions and must be consistent with the information in the product's approved label.

If a regulatory agency discovers previously unknown problems with our 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 or product candidate, including adverse events of unanticipated severity or frequency, or with our manufacturing processes, or fail to comply with regulatory requirements, a regulatory agency or enforcement authority may, among other things:

- issue warning letters;
- impose civil or criminal penalties;
- suspend or withdraw regulatory approval;

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

Any government investigation of alleged violations of law could require us to expend significant time, cost 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, co-marketers or other third-parties operating on our behalf fails to comply with regulatory requirements, regulatory authorities could impose fines on us, instate restrictions on our product or its manufacture or require us to recall or remove the product from the market, in addition to withdrawing our marketing authorizations, or requiring 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 such 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. 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 be subject to enforcement actions and we may not achieve or sustain corporate 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 and other healthcare professional payment and price transparency, 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.

Healthcare professionals, physicians and third-party payors play a primary role in the recommendation and prescription of ARCALYST and any product candidates for which we obtain marketing approval. Our commercial arrangements 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 ARCALYST and our product candidates for which we obtain marketing approval.

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

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

- the U.S. federal False Claims Act and civil monetary penalties laws, which, among other things, impose criminal and civil penalties, including through civil whistleblower or qui tam actions, against individuals or entities for knowingly presenting, or causing to be presented, to the federal government, claims for payment that are false or fraudulent or making a false statement to avoid, decrease or conceal an obligation to pay money to the federal government. 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. Moreover, 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;

- the U.S. Foreign Corrupt Practices Act, or FCPA, which prohibits U.S. companies and their representatives from paying, offering to pay, promising to pay or authorizing the payment of anything of value to any foreign government official, government staff member, political party or political candidate for the purpose of obtaining or retaining business or to otherwise obtain favorable treatment or influence a person working in an official capacity abroad. In many countries, the healthcare professionals we interact with may meet the FCPA's definition of a foreign government official. The FCPA also requires public companies to make and keep books and records that accurately and fairly reflect their transactions and to devise and maintain an adequate system of internal accounting controls;

- HIPAA, which imposes criminal and civil liability for executing a scheme to defraud any healthcare benefit program or obtain, by means of false or fraudulent pretenses, representations or promises, any of the money or property owned by, or under the custody or control of, any healthcare benefit program, regardless of the payor (e.g., public or private) and knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false statement in connection with the delivery of or payment for healthcare benefits, items or service. Similar to the federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation;

- the U.S. federal physician payment transparency requirements, sometimes referred to as the "Sunshine Act", which requires manufacturers of drugs, devices, biologics and medical supplies that are reimbursable under Medicare, Medicaid or the Children's Health Insurance Program to report to the Department of Health and Human Services information related to certain financial interactions with physicians (defined to include doctors, dentists, optometrists, podiatrists and chiropractors), certain non-physician practitioners including physician assistants and nurse practitioners, and teaching hospitals, as well as the ownership and investment interests of physicians and their immediate family members;

- analogous state laws and regulations, such as state antikickback and false claims laws that may apply to sales or marketing arrangements and claims involving healthcare items or services reimbursed by nongovernmental 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 professionals or marketing expenditures and pricing information; and

- similar healthcare laws and regulations in the EU and other jurisdictions, including reporting requirements detailing interactions with and payments to healthcare providers.

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, prescribers or other potential purchasers of our products or product candidates, if approved. We have entered into consulting and advisory board agreements with physicians and other healthcare professionals and could be adversely affected if regulatory authorities determine our financial relationships with such prescribers violate applicable laws or create a conflict of interest. For example, 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. Regulatory authorities may conclude that a financial relationship between us and a principal investigator or a clinical trial site has created a conflict of interest or otherwise affected interpretation of a study. Regulatory authorities may therefore question the integrity of the data generated at the applicable clinical trial site and the utility of the clinical trial itself may be jeopardized, which could result in a delay in approval, or rejection, of our marketing applications by regulatory authorities and may ultimately lead to the denial of marketing approval of our product candidates. Furthermore, investigators for our clinical trials may become debarred by regulatory authorities, which may impact the integrity of our studies and the utility of the clinical trial itself may be jeopardized. Because of the breadth of these laws and the narrowness of the statutory exceptions and regulatory safe harbors available under such laws, it is possible that some of our business activities could be subject to challenge under one or more of such laws. The scope and enforcement of each of these laws is uncertain and subject to rapid change in the current environment of healthcare reform, especially in light of the lack of applicable precedent and regulations.

Interactions between biopharmaceutical companies and healthcare professionals are also governed by strict laws, regulations, industry self-regulation codes of conduct and healthcare professionals' codes of professional conduct. The provision of any inducements to healthcare professionals to prescribe, recommend, endorse, order, purchase, supply, use or administer a drug product is prohibited. A number of countries have established additional rules requiring pharmaceutical companies to publicly disclose their interactions with physicians and other healthcare professionals and to obtain approval from employers, professional organizations or competent authorities before entering into agreements with healthcare professionals.

Ensuring that our business arrangements with third parties comply with applicable healthcare laws and regulations may 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 activities conducted by our sales team, were to be found to be in violation of any of these laws or any other governmental regulations that may apply to us, we may be subject to the imposition of civil, criminal and administrative penalties, damages, disgorgement, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, individual imprisonment, contractual damages, reputational harm, diminished profits and future earnings, additional reporting requirements or oversight if we become subject to a corporate integrity agreement or similar agreement to resolve allegations of non-compliance with these laws, and curtailment or restructuring of our operations, any of which could adversely affect our ability to operate our business and our results of operations. Further, defending against any such actions can be costly, time consuming and may require significant personnel resources. Therefore, even if we are successful in defending against any such actions that may be brought against us, our business may be impaired.

Risks Related to Product Development

We depend heavily on the success of one or more of our products and product candidates, which are in various stages of clinical development. If we are unable to advance our product candidates in clinical development, obtain regulatory approval and ultimately successfully commercialize one or more of our product candidates, or experience significant delays in doing so, our business will be significantly harmed.

We are not permitted to market or promote any of our product candidates before we receive regulatory approval from the FDA or comparable regulatory authorities outside of the United States. Our product candidates are in various stages of clinical development. Our assumptions about why our product candidates are worthy of future development and potential approval in the indications for which we are studying them, or any other indications, are based in part on indirect data collected by other companies and in part from data collected from our preclinical and clinical trials. We may not be able to demonstrate that they are safe or effective in the indications for which we are studying them, and they may not be approved.

We cannot be certain that any of our product candidates will be successful in their clinical trials or will receive regulatory approval even after completing a successful pivotal clinical trial. We may also determine that the potential product and commercial profile of any of our product candidates may not ultimately be commercially successful, or even if we believe they have the potential to be commercially successful, we may discontinue development of one or more of our product candidates or discontinue development in a specific indication for a particular product candidate, at any time

for any reason. If we do not receive regulatory approvals for more than one of our product candidates, we may not be able to continue our operations.

While we received FDA approval of ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older, each of our product candidates requires substantial additional preclinical or clinical development and manufacturing support and, if approved, an organization to facilitate a successful product launch and commercialization before we will be able to generate any revenue from product sales. The success of our product candidates or potential future product candidates depends upon several factors, including the following:

- submission to and authorization to proceed with clinical trials by the FDA under investigational new drug applications, or INDs, and clinical trial applications to applicable authorities outside of the United States for our product candidates to commence planned clinical trials or future clinical trials;
- successful completion of nonclinical studies, including toxicology studies, pharmacological, and biodistribution studies , as conducted, where applicable, under the FDA's good laboratory practices, or GLP regulations;
- successful site activation for, enrollment in, and completion of clinical trials, including the ability of our CROs to successfully conduct such trials within our planned budget and timing parameters without materially adversely impacting our trials, and our ability to successfully oversee CRO activities;
- positive data from our clinical programs, including post-marketing trials and those intended to satisfy regulatory commitments or for label expansion, with sufficient quality that support an acceptable risk-benefit profile of our products and product candidates for the targeted indications in the intended populations to the satisfaction of the applicable regulatory authorities;
- timely receipt, if at all, of approvals from applicable regulatory authorities and maintenance of any such approvals;
- as applicable, acceptance of pediatric study plans by regulatory authorities, and the follow through of any pediatric study commitments, such as development of pediatric formulations, if required;
- establishment and maintenance of arrangements with third party manufacturers, as applicable, for continued clinical supply and commercial manufacturing;
- successful development of our manufacturing processes and transfer to third party contract development and manufacturing organization, or CDMO, facilities to support our development and commercialization activities in a manner compliant with all regulatory requirements;
- successful manufacture of sufficient supply of our product candidates within approved specifications for purity and efficacy from our facility and from our CDMOs or other sole-source manufacturers in order to meet clinical or commercial demand, as applicable, for ourselves and for our partners;
- establishment and maintenance of patent and trade secret protection or regulatory exclusivity for our product candidates;
- timely and successful commercial launch of our product candidates;
- adoption of our products, if and when approved, by patients, patient-advocates, the medical community and third-party payors;
- effective competition with other therapies;

- establishment and maintenance of adequate healthcare coverage and reimbursement;
- enforcement and defense of intellectual property rights and claims;
- continued compliance with any postmarketing requirements imposed by regulatory authorities, including any required postmarketing clinical trial commitments or REMS or similar risk management measures; and
- maintenance of a continued acceptable safety profile of our product candidates following approval.

If we do not accomplish one or more of these factors in a timely manner or at all, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic, we could experience significant delays in, or an inability to, timely or successfully commercialize our product candidates, which would materially harm our business. If we do not receive regulatory approvals for one or more additional product candidates, we may not be able to continue our operations. Failure to generate sufficient revenue from the commercialization of our current and future products, whether as a result of failing to obtain regulatory approvals or unsuccessfully commercializing such products would harm our ability to continue our operations. In such an instance, we may need to seek capital elsewhere. See "*Risk Factors – Risks Related to Our Financial Position and Capital Needs – We will require substantial additional financing, and a failure to obtain this necessary capital when needed on acceptable terms, or at all, will force us to delay, limit, reduce or cease one or more of our product development plans, research and development programs for our product candidates, or other operations or commercialization efforts"* and "*Risk Factors – Risks Related to Our Financial Position and Capital Needs – Raising additional capital may cause dilution to our shareholders, restrict our operations or require us to relinquish rights to our technologies or product candidates or products."*

Even though we received FDA approval for ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older, and assumed the sales and distribution of ARCALYST for the previously approved indications in the United States, we evenly split profits on ARCALYST sales with Regeneron, and the relevant markets for these indications may prove not to be large enough to allow us to generate significant and sustained revenue from these product sales. Moreover, even if we successfully obtain regulatory approvals to manufacture and market one or more 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, among other things. If the markets for patient subsets that we are targeting are smaller than we estimate, we may not generate projected revenue levels from sales of such product candidates, if approved.

Clinical drug development is a lengthy and expensive process with uncertain timelines and 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 successfully commercialize our product candidates on a timely basis, if at all.

Before obtaining marketing approval from regulatory authorities for the sale of our product candidates, we must conduct extensive clinical trials to demonstrate the safety and efficacy of our product candidates in humans. Clinical testing is expensive, time consuming and uncertain as to outcome.

Not all of our clinical trials have been conducted as initially planned or completed on our initial projected schedule, and accordingly, we cannot guarantee that any of our current or potential future clinical trials will be conducted as initially planned or completed on our initial projected schedule, if at all, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic. For example, in December 2021, we announced that the primary efficacy endpoint of the Phase 3 clinical trial of mavrilimumab in COVID-19-related ARDS did not reach statistical significance. We subsequently decided to not progress mavrilimumab in the COVID-19-related ARDS indication. Clinical trials are a lengthy process that require the expenditure of significant money and human capital. Failing to achieve desired efficacy or identifying of a novel safety hazard in turn represents an inability to successfully recoup such expense via a potential commercialization of the product candidate, if approved. Sufficient inability to recoup clinical trial expense via successful development could pose material risks to our business. See "*Risk Factors – Risks Related to Product Development –We depend heavily on the success of one or more of our products and product candidates, which are in various stages of clinical development. If we are unable to advance our product candidates in*

clinical development, obtain regulatory approval and ultimately successfully commercialize one or more of our product candidates, or experience significant delays in doing so, our business will be significantly harmed."

Commencing a clinical trial is subject to acceptance by the FDA of an IND or IND amendments, acceptance by competent authorities of the EU member states of a clinical trial application, or CTA, or acceptance by other applicable regulatory authorities, and finalizing the trial design based on discussions with the FDA, competent authorities of the EU member states or other applicable regulatory authorities. We have and may in the future receive feedback or guidance from regulatory authorities on our clinical trial design and protocols and, even after we incorporate such feedback or guidance from these regulatory authorities, such regulatory authorities may impose other requirements for our clinical trials, could disagree that we have satisfied their requirements to commence our clinical trials, disagree with our interpretation of data from the relevant preclinical studies, clinical trials or chemistry, chemistry, manufacturing and controls, or CMC, data, or disagree or change their position on the acceptability of our trial designs, including the proposed dosing level or schedule, treatment duration, our definitions of the patient populations or the clinical endpoints selected, which may require us to complete additional preclinical studies, clinical trials, CMC development, other studies or impose stricter approval conditions than we currently expect.

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

Successful completion of our clinical trials is a prerequisite to submitting a BLA or supplemental BLA, or sBLA, to the FDA and an MAA, to the European Medicines Agency, or EMA, or competent authorities of the EU member states, or other applicable regulatory authorities in other countries for each product candidate and, consequently, to obtaining approval and initiating commercial marketing of our current and any future product candidates. A failure of one or more of our current or future clinical trials can occur at any stage of testing, and our clinical trials may not be successful. We have experienced and may continue to experience delays in our ongoing clinical trials, and we do not know whether planned clinical trials will begin on time, be allowed by regulatory authorities, need to be redesigned, or if we can activate sites or enroll patients on time, or if they will be completed on schedule, if at all. Events that have and may in the future delay or prevent commencement or successful completion of clinical development of our product candidates as planned and on schedule, if at all, include but are not limited to:

- inability to generate sufficient preclinical, toxicology or other in vivo or in vitro data to support the initiation of human clinical trials;
- delays or failure in reaching a consensus with regulatory agencies on trial design or implementation, including the appropriate dosage levels, frequency of dosing, or treatment period in clinical trials;
- delays or failure in reaching agreement on acceptable terms with prospective CROs, and clinical trial sites, the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and clinical trial sites;
- difficulties in obtaining required IRB or ethics committee approval at each clinical trial site;
- delays or failure in obtaining regulatory approval to commence a trial, or imposition of a clinical hold by regulatory authorities;
- difficulty in identifying and enrolling suitable patients in a particular trial, which may reduce the power of a clinical trial to detect statistically significant results;

- amendments to clinical trial protocols impacting study criteria, endpoints or design, including amendments that either we initiate or are requested by regulatory authorities;
- difficulty collaborating with patient groups and investigators;
- failure by our CROs, medical institutions, or other third parties we contract with in connection with our clinical trials to adhere to clinical trial requirements or to perform their obligations in a timely or manner compliant with all applicable laws and regulations, including FCPA;
- failure to perform in accordance with the FDA's good clinical practices, or GCPs, or applicable comparable regulatory guidelines in other countries;
- patients not completing a clinical trial or not returning for post-treatment follow-up, in either case including as a result of trial demands on participants as a result of the COVID-19 pandemic and measures taken in response to the pandemic or otherwise, among other things;
- clinical trial sites withdrawing from or being unable to conduct activities, or patients withdrawing from clinical trials, including as a result of the COVID-19 pandemic or the ongoing war in Ukraine, among other things;
- participating patients experiencing serious adverse events or undesirable side effects or being exposed to unacceptable health risks;
- participating patients failing to experience confirmed pre-specified events during the clinical trial within an expected timeframe, if at all;
- safety issues, including occurrence of adverse events associated with a product candidate, that are viewed to outweigh its potential benefits;
- changes in regulatory requirements, policies and guidance that require amending or submitting new clinical protocols;
- the cost of clinical trials being greater than we anticipate;
- strategic decisions regarding clinical study priority for capital preservation purposes;
- failure by us, our CROs, or other third parties with whom we contract to properly collect, analyze, and/or assess clinical data, including the performance of assays, analyses and other activities;
- clinical trials of our product candidates producing negative, inconclusive or uncompetitive results, which may result in us deciding, or regulatory authorities requiring us, to conduct additional clinical trials or modify or cease development programs for our product candidates;
- failure to replicate safety, efficacy or other data from earlier preclinical studies and clinical trials conducted by us or third parties, including the companies from whom we have licensed or acquired or may in the future license or acquire our product candidates, in our later clinical trials;
- the occurrence of adverse or other events not observed in earlier studies;
- suspensions or terminations of our clinical trials by us or the IRBs of the institutions in which our clinical trials are being conducted, the Data Safety Monitoring Board for such trials or the FDA or comparable regulatory authorities;

- failure of manufacturers, or us, to produce phase-appropriate supplies of our product candidates for use in our clinical trials in accordance with cGMP requirements and regulations or applicable comparable regulatory guidelines in other countries;

- delays in manufacturing, testing, releasing, validating or importing/exporting sufficient stable quantities of our product candidates for use in clinical trials or the inability to do any of the foregoing either as a result of quality assurance or due to our reliance on third party manufacturers; and

- disruptions to our business operations, including our manufacturing operations, and the business operations of our third-party manufacturers, CROs upon whom we rely to conduct our clinical trials, or other third parties with whom we conduct business or otherwise engage, as well as disruptions in supply chain distribution and business travel into and within the countries in which we conduct our clinical trials, our manufacturers produce our product candidates or we otherwise conduct business or engage with other third parties, now or in the future as a result of the impact of the COVID-19 pandemic.

Delays in the commencement or completion of our planned and ongoing clinical trials of our product candidates have occurred and may continue to occur. Consequences of delays have increased and may in the future increase our costs of developing our product candidates, slow down the development and approval of our product candidates, delay or jeopardize our ability to commence product sales and generate revenue, if any, from our product candidates and harm their commercial prospects. Furthermore, disruptions caused by the COVID-19 pandemic have increased and may continue to increase the likelihood that we encounter such difficulties or delays in commencing or completing our planned and ongoing clinical trials or other development. In addition, many of the factors that cause, or lead to, difficulties and delays in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates or us deciding to modify or cease development of our product candidates.

Clinical trial delays could also shorten any periods during which our products have patent protection or shorten any periods during which we have the exclusive right to commercialize our product candidates and may allow our competitors to bring products to market before we do, which could impair our ability to obtain orphan exclusivity for our products that potentially qualify for this designation and to successfully commercialize our product candidates, and may harm our business and results of operations. Any inability to successfully complete preclinical and clinical development could result in additional costs to us or impair our ability to generate revenue and harm our business, financial condition and prospects significantly.

Furthermore, clinical trials must be conducted in accordance with the laws, rules and regulations, guidelines and other requirements of the FDA, EU institutions, the European Medicines Agency, or EMA and other applicable regulatory authorities outside of those jurisdictions and are subject to oversight by these regulatory authorities and IRBs or ethics committees at the medical institutions where the clinical trials are conducted. Further, conducting global clinical trials, as we do for certain of our product candidates, may require that we coordinate among the legal requirements and guidelines of regulatory authorities across a number of jurisdictions, including the United States, EU and countries outside of those jurisdictions, which could require that we amend clinical trial protocols or determine not to conduct a trial in one or more jurisdictions or to run separate trials in various jurisdictions due to the inability, cost or delay in harmonizing divergent requests from such regulatory authorities, all of which could increase costs. In addition, clinical trials that are conducted in countries outside the United States and the EU may subject us to risks associated with the engagement of non-United States and non-EU CROs who are unknown to the FDA or the EMA, or the EU member states' regulatory authorities and may have different standards of diagnosis, screening and medical care, as well as risks associated with further delays and expenses as a result of increased shipment costs (including as a result of local quality release or in-country testing of a product candidate supply produced in a different jurisdiction for our clinical trials) and political and economic risks relevant to such countries outside the United States and the EU.

In addition, the FDA's and other regulatory authorities' policies with respect to clinical trials may change and additional government regulations may be enacted. For instance, the regulatory landscape related to clinical trials in the EU recently evolved. The EU CTR, which was adopted in April 2014 and repeals the EU Clinical Trials Directive, became applicable on January 31, 2022. While the Clinical Trials Directive required a separate CTA to be submitted in each member state, to both the competent national health authority and an independent ethics committee, the CTR

introduces a centralized process and only requires the submission of a single application to all member states concerned. The CTR allows sponsors to make a single submission to both the competent authority and an ethics committee in each member state, leading to a single decision per member state. The assessment procedure of the CTA has been harmonized as well, including a joint assessment by all member states concerned, and a separate assessment by each member state with respect to specific requirements related to its own territory, including ethics rules. Each member state's decision is communicated to the sponsor via the centralized EU portal. Once the CTA is approved, clinical study development may proceed. The CTR foresees a three-year transition period. The extent to which ongoing and new clinical trials will be governed by the CTR varies. For clinical trials whose CTA was made under the Clinical Trials Directive before January 31, 2022, the Clinical Trials Directive will continue to apply on a transitional basis for three years. Additionally, sponsors may still choose to submit a CTA under either the Clinical Trials Directive or the CTR until January 31, 2023 and, if authorized, those will be governed by the Clinical Trials Directive until January 31, 2025. By that date, all ongoing trials will become subject to the provisions of the CTR.

It is currently unclear to what extent the United Kingdom, or UK will seek to align its regulations with the EU. The UK regulatory framework in relation to clinical trials is derived from existing EU legislation (as implemented into UK law, through secondary legislation).

If we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies governing clinical trials, our development plans may be impacted.

Further, the ongoing war in Ukraine may also materially affect our clinical activities and our product candidate development timeline. See "*Risk Factors – General Risk Factors – The ongoing war in Ukraine, and actions taken against Russia as a result of its invasion of Ukraine, has and may continue to have an adverse impact on the global economy, equity capital markets and our clinical operations*."

The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact on our current or planned preclinical studies and clinical trials, which could be significant.

The COVID-19 pandemic, and measures taken in response to the pandemic, have had and could continue to have an impact on our current or planned preclinical studies and clinical trials. If the COVID-19 pandemic and measures undertaken in response to the pandemic are reinstated, including as a result of the emergence of new variants of the virus, we may experience significant disruptions that could materially impact our preclinical studies and clinical trials, including by:

- impeding, delaying, limiting or preventing the production, delivery or release of our product candidates and important ancillary products to our clinical trial sites or patients, including due to interruptions in the supply of raw materials or global shipping that may affect the transport of our product candidates or clinical trial materials, or the reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19;

- impeding, delaying, limiting or preventing the production, delivery or release of the supply of our product candidates, including due to disruptions at manufacturing facilities that produce our product candidates, staffing shortages, reprioritizations, production slowdowns or stoppages or interruptions in global shipping;

- impeding, delaying, limiting or preventing clinical trial investigators, other critical staff, or patients from traveling to our clinical trial sites or visiting nurses traveling to patients;

- impeding, delaying, limiting or preventing key clinical trial activities, including patient screening, clinical trial site monitoring, patient dosing, study procedures (such as biopsies, which may be deemed non-essential), collection of clinical data and samples as well as cleaning and verification of clinical data, which could affect the integrity of clinical trial data;

- diverting healthcare resources away from the conduct of clinical trials or reprioritizing the focus of such resources on clinical trials for product candidates with the potential for treatment or prevention of COVID-19 related conditions;

- timing of COVID-19 and other vaccinations received by potential patients for our clinical trials may impede, delay, limit or prevent such potential patients from enrolling in our clinical trials;

- impeding, delaying, limiting or preventing clinical trial site initiation, including difficulties in recruiting clinical site investigators and clinical site staff, and enrollment or retention of patients in our clinical trials;

- increasing the risk that participants enrolled in our clinical trials will contract COVID-19 while the clinical trial is ongoing, which could impact the results of the clinical trial, including by increasing the number of observed adverse events;

- interrupting or delaying preclinical studies due to restricted or limited operations at our research and development laboratory facility;

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

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

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

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

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

Any one of the foregoing could significantly impede, delay, limit or prevent the clinical development of our product candidates and ultimately lead to the delay or denial of regulatory approval of our product candidates. While we continuously look to identify business-critical activities and to develop contingencies and mitigation strategies for those activities to potentially minimize the impact of the COVID-19 pandemic, or any other pandemic, on our business and operations, there can be no assurance that we will be able to identify all such activities or that any identified contingencies and mitigation strategies will be effective. If the clinical development of our product candidates is significantly impeded, delayed, limited or is prevented, it could ultimately lead to the delay or denial of regulatory approval of our product candidates which would materially adversely affect our business and operations, including our ability to generate revenue.

We may find it difficult to enroll patients in our clinical trials in a timely manner given the limited number of patients who have the diseases for which our product candidates are being studied, our particular enrollment criteria or competing clinical studies in the same patient population.

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

patients to participate in testing our product candidates, particularly given that many of the conditions for which we are evaluating our current product candidates or may evaluate them in the future are in small disease populations. In addition, the eligibility criteria of our clinical trials will further limit the pool of available trial participants, as we will require patients to have specific characteristics that we can evaluate based on the primary and secondary endpoints of the study. Further, our product candidates modulate the immune system and carry risks associated with immunosuppression, including the risk of serious infections, potential interference with vaccines, and other potential serious health risks. Additionally, certain indications for our product candidates may present challenges that may prevent us or third parties from conducting well-controlled studies.

Our clinical trials have competed and may continue to compete with other clinical trials for product candidates that are in the same therapeutic areas as our product candidates. This competition may further reduce the number and types of patients available to us because some patients who might have opted to enroll in our trials may instead opt to enroll in a trial being conducted by one of our competitors. Since the number of qualified clinical investigators is limited, we may conduct some of our clinical trials at the same clinical trial sites that some of our competitors use, which would reduce the number of patients who are available for our clinical trials at such clinical trial site.

Accordingly, when we encounter these or other difficulties in enrollment, we have experienced and may continue to experience delays or we may be prevented from completing our clinical trials. Patient enrollment depends on many factors, including:

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

The process of finding and enrolling patients may prove costly, especially since we are looking to identify a subset of the patients eligible for our studies from a relatively small patient population for many of the diseases we are studying. If patients are unable or unwilling to participate in our clinical trials for any reason, or we experience

difficulties in patient enrollment for any other reasons, such as due to the COVID-19 pandemic, our costs may significantly increase and the timeline for recruiting patients, conducting trials and obtaining regulatory approval of our product candidates may be significantly delayed or prevented, the commercial prospects of our product candidates may be harmed, and our ability to commence product sales and generate product revenue from any of these product candidates, if approved, could be delayed or prevented. Any of these occurrences may harm our business, financial condition, and prospects significantly.

Our products and 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 products and product candidates may produce undesirable side effects or adverse reactions or events. Undesirable side effects caused by our product candidates could cause us or regulatory authorities to interrupt, delay or halt clinical trials and could result in more restrictive labels or the delay or denial of regulatory approvals by regulatory authorities.

Our products and product candidates modulate the immune system and carry risks associated with immunosuppression, including the risk of serious infections and other potential serious health risks.

For mavrilimumab, there is a theoretical risk for the development of pulmonary alveolar proteinosis, or PAP, with chronic use. PAP is a rare lung disorder in which surfactant-derived lipoproteins accumulate excessively within pulmonary alveoli due to loss of GM-CSF function. The disease can range in severity from a sub-clinical reduction in diffusion capacity to significant dyspnea during mild exertion. In preclinical studies conducted by MedImmune, certain effects were observed in the lungs of non-human primates, which led the FDA to issue a clinical hold with respect to MedImmune's proposed clinical trial in RA. Preclinical data generated to-date suggest mavrilimumab at clinically relevant doses does not reach the lungs in sufficient quantities to induce PAP, and human trials thus far have not shown a clinical effect on pulmonary function tests attributable to mavrilimumab.

However, if the results of our clinical trials, or clinical trials conducted by collaboration partners, reveal an unacceptable severity and prevalence of certain side effects, the FDA or applicable regulatory authority outside of the United States may suspend or terminate our clinical trials, or not authorize us to initiate further trials. In addition, if other molecules in the same or related class in development by third parties show the same or similar side effects as those we observed in our trials but to a greater degree or reported new previously unreported side effects, it could have an impact on the entire class of molecules in development, as the applicable regulatory agency may suspend or terminate our clinical trials, or not authorize us to initiate further trials with our molecule in that class. Such effects, if discovered by third-parties, may not be discovered until after we commence the commercialization of a product. Regulatory authorities could order us to cease further development of, deny or withdraw any approval of, any of our products or product candidates, or require onerous label changes, for any or all targeted indications in such an instance.

In addition, the compassionate use of our product candidates, or evaluation of our product candidates by third parties via scientific collaborations or investigator initiated studies could increase the possibility of generating adverse safety results that impact our development of such product candidates. Such adverse safety results, when reported to regulatory authorities, may negatively impact the safety profile of the drug studied as a class effect and could result in the imposition of clinical holds on all clinical trials involving such product candidate regardless of the indication studied.

Further, clinical trials by their nature utilize a sample of the potential patient population. Certain rare and severe side effects associated with our products or product candidates may only be uncovered with a significantly larger number of patients exposed to the product candidates. If we or others later identify undesirable side effects caused by our product or any of our product candidates, if approved, a number of potentially significant negative consequences could result, including but not limited to:

- regulatory authorities may withdraw approvals of such product and require us to take it off the market;

- regulatory authorities may require the addition of labeling statements, specific warnings, contraindications or field alerts to prescribers and pharmacies;
- we may be required to create a registry or a REMS plan or similar risk management measures, which could include a medication guide outlining the risks of such side effects for distribution to patients, a communication plan for healthcare providers or other elements to assure safe use;
- we may be required to change the way the product is administered, conduct additional clinical trials or change the labeling of the product;
- we may be subject to limitations on how we promote the product, or sales of the product may decrease significantly;
- we could be sued and held liable for harm caused to patients;
- the product may become less competitive; and
- our reputation may suffer.

Any of these events could prevent us from achieving or maintaining market acceptance of the particular product or product candidate, if approved, and could significantly harm our business, results of operations and prospects.

Interim, preliminary, and "top-line" data from our clinical trials that we announce or publish from time to time may change as more patient data become available following the release of the interim data; preliminary data are subject to audit and verification procedures, and deeper analysis of the data beyond the topline data may provide more color and context to the data, all of which could result in material or other changes that are reflected in the final data.

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

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

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

If the interim, preliminary, or topline data that we report differ materially from final results, or if third parties, including regulatory authorities, disagree with the conclusions reached, our ability to obtain approval for, and

commercialize, our product candidates may be harmed, which could harm our business prospects, operating results or financial condition. Further, announcement of preliminary, interim or top-line data by us or differences between that data and the final data could result in volatility in the price of our Class A common shares.

Risks Related to Marketing Approval and Regulatory Matters

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

Before we can commercialize any of our current or future product candidates, we must obtain marketing approval from regulatory authorities. We may not be able to receive approval to market any of our current or future product candidates from regulatory authorities in our desired indications 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. We may need to rely on third party CROs and regulatory consultants to assist us in this process. Securing regulatory approval requires the submission of extensive preclinical and clinical data and supporting information to the various regulatory authorities for each therapeutic indication to establish a product candidate's safety and efficacy. Securing regulatory approval also requires the submission of information about the biologic manufacturing process to, and inspection of manufacturing facilities by, the relevant regulatory authorities, who may deny approval based on the results of such submissions and inspections. Our current or future 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, including 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 or such authorities may request additional information that may be difficult to generate or provide. Further, following approval, the FDA may conduct additional inspections and, based on the results of such inspections, deem the inspected manufacturing facilities to be deficient, suspending our ability to manufacture our product candidates until we can secure satisfactory alternative manufacturing facilities.

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

The process of obtaining regulatory approvals, both in the United States and in other countries, is time consuming, expensive, may take many years, if approval is obtained at all, and can vary substantially based upon a variety of factors, including the type, complexity and novelty of the product candidates involved. Changes in marketing approval policies during the development period, changes in or the enactment of additional statutes or regulations, or changes in regulatory review for each submitted BLA, or equivalent application types, may cause delays in the approval or rejection of an application. For instance, the EU pharmaceutical legislation is currently undergoing a complete review process, in the context of the Pharmaceutical Strategy for Europe initiative, launched by the European Commission in November 2020. A proposal for revision of several legislative instruments related to medicinal products (potentially revising the duration of regulatory exclusivity, eligibility for expedited pathways, etc.) is expected to be adopted by the European Commission between the end of 2022 and early Q2 of 2023. The proposed revisions, once they are agreed and adopted by the European Parliament and European Council (not expected before the end of 2024), may have a significant impact on the biopharmaceutical industry in the long term.

Regulatory authorities have substantial discretion in the approval process and may refuse to accept any application or may decide that our data are insufficient for approval and require additional preclinical studies or clinical or other trials for our current or future product candidates. Our current and future product candidates could be delayed in

receiving, or fail to receive, regulatory approval or we may fail or cease to advance their development for many reasons, including the following:

- 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 regulatory authorities that a product candidate is safe and effective for its proposed indication or that its clinical and other benefits outweigh its safety risks;
- regulatory authorities could require us to collect additional data or conduct additional clinical trials, which could include a requirement to compare our product candidates to other therapies for the treatment of the same indication;
- the results of clinical trials may produce negative, inconclusive or uncompetitive results, which may result in us deciding, or regulatory authorities requiring us, to conduct additional clinical trials or to modify or cease development programs for our product candidates;
- the results of clinical trials may not meet the primary or secondary endpoints of the applicable trial or the level of statistical significance required by regulatory authorities;
- regulatory authorities may disagree with our interpretation of data from preclinical studies or clinical trials;
- the data collected from clinical trials of our product candidates may not be sufficient to support the submission of a BLA, sBLA or other submission or to obtain regulatory approval in the United States or elsewhere;
- the 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 data quality and regulatory requirements or meet their contractual obligations to us in a timely manner, or at all;
- regulatory authorities may not believe that we have sufficiently demonstrated our ability to manufacture the products to the requisite level of quality standards, including that such material is sufficiently comparable to material used in previous clinical trials, 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;
- regulatory authorities may not believe that their on-site inspections and data audits have sufficiently demonstrated the quality and integrity of the clinical trial conduct and of data submitted to regulatory authorities in support of our new product approvals and marketing applications;
- the supply or quality of our product candidates or other materials necessary to conduct clinical trials of our product candidates may be insufficient or inadequate;
- our product candidates may have undesirable side effects, toxicities or other unexpected characteristics, causing us or our investigators, regulatory authorities or IRBs to reject, suspend or terminate the clinical trials; and

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

In addition, even if we were to obtain approval for one or more of our current or future product candidates, regulatory authorities may approve any of our current or future product candidates for fewer indications or more limited patient populations than we request. Furthermore, regulatory authorities may not approve the price we intend to charge, may grant approval contingent on the performance of costly post-marketing clinical trials, may impose certain post-marketing requirements that impose limits on our marketing and distribution activities, or may approve a product candidate with a label that does not include the labeling claims necessary or desirable for the successful commercialization of that product candidate. Any of the foregoing scenarios could materially harm the commercial prospects for our current or future product candidates.

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

Our products, current product candidates and any of our future product candidates regulated as biologics in the United States may face biosimilar competition sooner than anticipated.

In the United States, the BPCIA created an abbreviated approval pathway for biological products that are biosimilar to or interchangeable with an FDA licensed reference biological product. Under the BPCIA, an application for a biosimilar product may not be submitted to the FDA until four years following the date that the reference product was first approved under a BLA by the FDA. In addition, the approval of a biosimilar product may not be made effective by the FDA until 12 years from the date on which the reference product was first licensed. During this 12 year period of exclusivity, another company may still market a competing version of the reference product for the same therapeutic indication if the FDA approves a full BLA for the competing product containing the sponsor's own preclinical data and data from adequate and well controlled clinical trials to demonstrate the safety, purity and potency of their product. The law is complex and is still being interpreted and implemented by the FDA. As a result, its ultimate impact, implementation and meaning are subject to uncertainty.

For example, although ARCALYST was approved as a biological product under a BLA for the treatment of CAPS in February 2008, and we believe it qualified for the 12-year period of exclusivity against any biosimilars, such 12-year period of exclusivity has lapsed. The FDA approved ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021. However, the 12-year exclusivity period does not attach to the approval of an sBLA, potentially creating the opportunity for biosimilar competition, subject to any Orphan Drug exclusivity under the U.S. Orphan Drug Act (See "*Risk Factors — Risks Related to Marketing Approval and Regulatory Matters — We may seek Orphan Drug designation for our product candidates in the United States, as well as for any of our product candidates in the EU, and we may be unsuccessful, or may be unable to maintain the benefits associated with Orphan Drug designation, including the potential for market exclusivity, for any product candidate for which we obtain Orphan Drug designation.")*. If we obtain FDA approval for any of our other biological product candidates, we expect any such product candidates to qualify for the 12-year period of exclusivity under the BPCIA. However, there is a risk that this exclusivity could be shortened due to Congressional action or otherwise, or that the FDA will not consider any such approved product candidates to be reference products for competing products, potentially creating the opportunity for generic competition sooner than anticipated.

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

In order to market any products in a country or territory, we must establish and comply with numerous and varying regulatory requirements of such country or territory regarding safety and efficacy. Regulatory requirements can vary widely from country to country, and clinical trials conducted in one country may not be accepted by regulatory authorities in other countries, and regulatory approval in one country does not mean that regulatory approval will be

obtained in any other country. Approval procedures vary among countries and can involve additional product testing and validation, additional administrative review periods, and additional preclinical studies or clinical trials, which would be costly and time consuming and could delay or prevent the introduction of our current or future product candidates, or ARCALYST, 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 our product candidates in the United States, as well as for any of our product candidates in the EU, and we may be unsuccessful, or may be unable to maintain the benefits associated with Orphan Drug designation, including the potential for market exclusivity, for any product candidate for which we obtain Orphan Drug designation.

We have received Orphan Drug designation in the United States for mavrilimumab for the treatment of GCA, and we may seek Orphan Drug designation for certain of our other product candidates in the United States as well as for any of our product candidates in the EU. We may be unsuccessful in obtaining such designation for any of our other product candidates or unable to maintain the associated benefits for any of our other current or future product candidates that are granted Orphan Drug designation, if any. Regulatory authorities in some jurisdictions, including the United States and Europe, may designate drugs or biologics intended to treat relatively small patient populations as Orphan Drug products. Under the U.S. Orphan Drug Act, the FDA may designate a drug or biologic as an Orphan Drug if it is intended to treat a rare disease or condition, which is generally defined as a patient population of fewer than 200,000 individuals in the United States, or a patient population greater than 200,000 in the United States where there is no reasonable expectation that the cost of developing the drug will be recovered from sales in the United States.

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

In addition, if a drug or biologic with an Orphan Drug designation subsequently receives the first marketing approval for the disease or condition for which it has such designation, the drug or biologic is entitled to a period of marketing exclusivity, which precludes a regulatory authority from approving another marketing application for the same drug and disease or condition 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 diseases or conditions as our product candidates, we may not be able to have competing products approved by the applicable regulatory authority for a significant period of time. The applicable period is seven years in the United States and ten years in the EU. The EU exclusivity period can be reduced to six years if, at the end of the fifth year, it is established that a drug no longer meets the criteria for Orphan Drug designation including where it is shown that the drug is sufficiently profitable not to justify maintenance of market exclusivity.

In connection with the FDA's approval of ARCALYST in the recurrent pericarditis indication, we received seven years of Orphan Drug exclusivity for ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients 12 years and older. 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 disease or condition. Even after an Orphan Drug is approved, the FDA can subsequently approve a later application for the same drug for the same disease or 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. Foreign regulatory authorities may also make the same determination. 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.

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

We may seek Breakthrough Therapy or Fast Track designation for one or more of our product candidates. A Breakthrough Therapy is defined as a drug or biologic that is intended, alone or in combination with one or more other drugs or biologics, to treat a serious or life-threatening disease or condition, and preliminary clinical evidence indicates that the drug or biologic may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. Separately, if a product candidate is intended for the treatment of a serious or life-threatening condition and clinical or preclinical data demonstrate the potential to address unmet medical needs for this condition, the sponsor may apply for Fast Track designation. Both Fast Track designation and Breakthrough Therapy designations offer sponsors the potential for rolling review of a BLA, where the FDA may consider for review sections of the BLA on a rolling basis before the complete application is submitted, if the sponsor provides a schedule for the submission of the sections of the BLA, the FDA agrees to accept sections of the BLA and determines that the schedule is acceptable, and the sponsor pays any required user fees upon submission of the first section of the BLA.

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

We may seek PRIME designation by EMA or other designations, schemes or tools in the EU, including the conditional marketing authorization or marketing authorization under exceptional circumstances, for one or more of our product candidates, which we may not receive. Such designations may not lead to a faster development or regulatory review or approval process and do not increase the likelihood that our product candidates will receive marketing authorization.

We may seek EMA PRIME (Priority Medicines) designation or other designations, schemes or tools for one or more of our product candidates. In the European Union, innovative products that target an unmet medical need and are expected to be of major public health interest may be eligible for a number of expedited development and review programs, such as the PRIME scheme, which provides incentives similar to the Breakthrough Therapy designation in the United States. PRIME is a voluntary scheme aimed at enhancing the EMA's support for the development of medicines that target unmet medical needs. It is based on increased interaction and early dialogue with companies developing promising medicines, to optimize their product development plans and speed up their evaluation to help them reach patients earlier. The benefits of a PRIME designation include the appointment of a rapporteur before submission of a marketing authorization application, early dialogue and scientific advice at key development milestones, and the potential to qualify products for accelerated review earlier in the application process.

Even if we believe one of our product candidates is eligible for PRIME, the EMA may disagree and instead determine not to make such designation. The EMA PRIME scheme or other schemes, designations, or tools, even if obtained or used for any of our product candidates may not lead to a faster development, regulatory review or approval process compared to therapies considered for approval under conventional procedures and do not assure ultimate

approval. In addition, even if one or more of our product candidates is eligible to the PRIME scheme, the EMA may later decide that such product candidates no longer meet the conditions for qualification or decide that the time period for review or approval will not be shortened.

Product developers that benefit from PRIME designation may be eligible for accelerated assessment (in 150 days instead of 210 days), which may be granted for medicinal products of major interest from a public health perspective or that target an unmet medical need, but this is not guaranteed.

Moreover, in the EU, a "conditional" marketing authorization may be granted in cases where all the required safety and efficacy data are not yet available. A conditional marketing authorization is subject to conditions to be fulfilled for generating missing data or ensuring increased safety measures. A conditional marketing authorization is valid for one year and has to be renewed annually until fulfillment of all relevant conditions. Once the applicable pending studies are provided, a conditional marketing authorization can become a "standard" marketing authorization. However, if the conditions are not fulfilled within the timeframe set by the EMA, the marketing authorization will cease to be renewed. Furthermore, marketing authorizations may also be granted "under exceptional circumstances" when the applicant can show that it is unable to provide comprehensive data on the efficacy and safety under normal conditions of use even after the product has been authorized and subject to the introduction of specific procedures. This may arise when the intended indications are very rare and, in the present state of scientific knowledge, it is not possible to provide comprehensive information, or when generating data may be contrary to generally accepted ethical principles. This type of marketing authorization is close to a conditional marketing authorization as it is reserved to medicinal products to be approved for severe diseases or unmet medical needs and the applicant does not hold the complete data set legally required for the grant of a marketing authorization. However, unlike a conditional marketing authorization, the applicant does not have to provide the missing data and will never have to. Although a marketing authorization "under exceptional circumstances" is granted definitively, the risk-benefit balance of the medicinal product is reviewed annually and the marketing authorization may be withdrawn where the risk-benefit ratio is no longer favorable.

The competent regulatory authorities in the EU have broad discretion whether to grant such an accelerated assessment, conditional marketing authorization or marketing authorization under exceptional circumstances, and, even if such assessment or authorization is granted, we may not experience a faster development process, review or authorization compared to conventional procedures. Moreover, the removal or threat of removal of such marketing authorizations may create uncertainty or delay in the clinical development of our product candidates and threaten the commercialization prospects of our products and product candidates, if approved. Such an occurrence could materially impact our business, financial condition and results of operations.

We have limited experience obtaining marketing approvals, and we may be unable to successfully do so for any of our current or future product candidates. Failure to successfully complete another pivotal clinical trial or obtain marketing approval in a timely manner for any of our current or future product candidates could have a material adverse impact on our business and financial performance.

Conducting pivotal clinical trials and preparing, and obtaining marketing approval for, a product candidate is a complicated process. As a company, we have only limited experience in obtaining marketing approval for our product candidates. As a result, in the future, obtaining marketing approval for any of our current or future product candidates may require more time and expense than we anticipate. Failure to successfully complete, or delays in, any of our eventual other pivotal trials or related regulatory submissions would prevent us from, or delay us in, obtaining regulatory approval for our current or future product candidates. It is possible that regulatory authorities may refuse to accept for substantive review any regulatory submissions that we submit for our product candidates or may conclude after review of our applications for any of our current or future product candidates that the submissions are insufficient to obtain marketing approval for such product candidates. Regulatory authorities may also require that we conduct additional clinical, preclinical or manufacturing validation trials and submit that data before they will reconsider our applications. Depending on the extent of these or any other required trials, approval or receipt of any marketing authorization may be delayed by several years or may require us to expend more resources than we have available. It is also possible that additional trials, if performed and completed, may not be considered sufficient by regulatory authorities to approve or grant marketing authorizations. Any delay in obtaining, or an inability to obtain, marketing approvals would delay or prevent us from commercializing any of our current or future product candidates, generating additional revenue and

achieving and sustaining corporate profitability. If any of these outcomes occur, we may be forced to modify or cease our development efforts for one or more of our product candidates, which could significantly harm our business.

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

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

Separately, in response to the COVID-19 pandemic, the FDA postponed inspections at facilities from March 2020 until July 2021. Even though the FDA has since resumed standard inspection operations of domestic facilities where feasible, it is nonetheless faced with a significant backlog as a result of its earlier suspension. Further, the FDA has continued to monitor and implement changes to its inspection activities to ensure the safety of its employees and those of the firms it regulates as it adapts to the evolving COVID-19 pandemic. Any resurgence of the virus may cause additional postponements, exacerbating the previously discussed risks.

Regulatory authorities outside the United States have adopted similar restrictions or other policy measures in response to the COVID-19 pandemic. If a prolonged government shutdown occurs, or if global health concerns continue to prevent regulatory authorities from conducting their regular inspections, reviews, or other regulatory activities, it could significantly impact the ability of such regulatory authorities to timely review and process our regulatory submissions, which could have a material adverse effect on our business.

Risks Related to Manufacturing and Our Reliance on Third Parties

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

We do not currently own or operate any late-stage or commercial manufacturing facilities. Although we have built a development and manufacturing facility to produce drug substance to support certain research, preclinical and other clinical development for our product candidates, we rely, and expect to continue to rely, on third parties for the manufacture of our late-stage product candidates and certain early-stage product candidates for the majority of our clinical development efforts, as well as for the commercial manufacture of ARCALYST or any of our current or future product candidates, if approved, as well as label and packaging activities. We rely on these third parties to produce ARCALYST and our product candidates at sufficient quality and quantity to support our commercialization and research and development efforts.

Our reliance increases the risk that we will have insufficient quantities of ARCALYST and our product candidates or that ARCALYST and our product candidates are not produced at an acceptable cost or quality, which

could delay, prevent or impair our commercialization or research and development efforts. For example, in the second half of 2021, events were identified in the ARCALYST manufacturing process that prevented distribution of certain ARCALYST material as previously planned. While we were ultimately able to source sufficient ARCALYST material to cover our current commercial needs, if we encounter events in the future that prevent additional material from being distributed in a timely manner or within specifications and we are unable to source additional commercial supply of ARCALYST, if needed, or should additional manufacturing or supply chain issues arise, we may be unable to adequately meet patient demand for ARCALYST or may be required to effect a recall, any of which would adversely affect our business, results of operations and financial condition. In addition, we may have difficulty sourcing an adequate supply of ARCALYST to cover our commercial and clinical needs, should we decide to develop ARCALYST in one or more additional indications or in additional territories.

Regeneron and its CMOs are the sole manufacturers of ARCALYST, and we have a contract with Regeneron to produce ARCALYST on an exclusive basis, subject to limited exceptions. However, Regeneron is not obligated to accept our forecasts or purchase orders that are not in line with accepted forecasts and Regeneron may not have sufficient manufacturing capacity to meet our commercial or clinical demand for ARCALYST. Regeneron, in turn, relies upon CMOs or other third parties to conduct fill/finish operations for ARCALYST. In the event that a particular batch of ARCALYST fails to meet specifications, whatever the cause, we are nonetheless obligated to pay for such material pursuant to the terms of the Supply Agreement. As a result of our reliance on Regeneron and its CMOs as our sole manufacturers, we do not have control over their manufacturing operations and scheduling, which may impact our ability to meet commercial or clinical demand for ARCALYST. We may also be subject to unexpected costs arising from any manufacturing or supply chain disruptions, which may materially impact our business, results of operations and financial condition.

Under certain circumstances, we or Regeneron could initiate a technology transfer to either us or another CMO to manufacture ARCALYST. For example, Regeneron could initiate a technology transfer as early as March 2023. Finding new CMOs or third-party suppliers to produce ARCALYST would add additional costs and require significant time and focus from our management and technical teams. We may also not be able to identify a suitable CMO that can meet our manufacturing timeline requirements. Even if we were to find a CMO, such CMO would need to produce ARCALYST at a different manufacturing site, potentially using a different or more costly process, or at a different scale. We cannot provide any assurance that the technology transfer from Regeneron to us or to another CMO will be successful in producing ARCALYST in sufficient quantities or of acceptable quality, if at all, or that we or another CMO will produce a comparable product to the satisfaction of regulatory authorities, which could delay, prevent or impair the further development, if any, or commercialization of ARCALYST. Further, we may be unable to establish a new agreement with another CMO on acceptable terms, if at all. In addition, there is typically a transition period when a new CMO commences work. Failure to produce ARCALYST in sufficient quantities or of acceptable quality could result in supply shortages for our patients, result in lost revenue and impact our ability to hold sufficient quantities of safety stock to be properly positioned to address unexpected disruptions to the ARCALYST supply chain.

We have entered into certain collaboration agreements with Huadong for each of ARCALYST and mavrilimumab. Until such time as Huadong is able to manufacture these products, either on its own or through a third-party CMO, we are the only source of these products for Huadong. If our current suppliers of drug substance and drug product for ARCALYST and mavrilimumab cannot produce sufficient quantities to satisfy our needs and Huadong's needs, then this may have an adverse impact on our and Huadong's business and operations.

In addition, we have engaged CMOs to manufacture mavrilimumab drug substance and drug product and have qualified CMOs to produce KPL-404 drug product. While we have manufacturing capabilities to support early development for our product candidates, we and our CMOs may not be able to produce sufficient quantities of our product candidates or produce them at an acceptable quality, including as a result of the COVID-19 pandemic or global supply chain issues, which could delay, prevent or impair our development or commercialization efforts and increase costs.

Our suppliers may also be negatively impacted by the COVID-19 pandemic. See "*Risk Factors — General Risk Factors — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and*

other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position."

If we make manufacturing or formulation changes to our products or product candidates or change manufacturers or manufacturing processes, we may be unsuccessful in producing products or product candidates comparable to those used in prior clinical trials. Therefore, we may need to conduct additional process development or additional clinical trials to bridge our prior clinical results to those resulting from the new manufacturing process, which could impact the timing and subsequent success of our planned clinical trials. In addition, as we plan to produce clinical trial and commercial material at a CMO, the CMO may be required to adopt different manufacturing protocols or processes. For example, although Regeneron has produced ARCALYST for commercial use for over ten years, regulatory authorities may reevaluate ARCALYST's current manufacturing processes or route of administration in connection with evaluating whether to approve ARCALYST for any new indication or for additional territories in the future or in connection with a technology transfer from Regeneron to us or another CMO.

The facilities used by our CMOs to manufacture ARCALYST and our current and future product candidates may be inspected by regulatory authorities in connection with the submission of our marketing applications to, and review by, regulatory authorities or based on their work for other clinical trial sponsors. While we provide oversight of manufacturing activities, we do not and will not control the manufacturing process of, and will be completely dependent on, our CMOs for compliance with cGMPs and other regulatory requirements in connection with the manufacture of ARCALYST and our product candidates. If our CMOs cannot successfully manufacture material that conforms to our specifications and the strict regulatory requirements of regulatory authorities, they will not be able to secure or maintain regulatory approval for their manufacturing facilities. While we review the compliance history and performance of our CMOs and have the ability to audit their compliance and performance, we have no direct control over the ability of our CMOs to maintain adequate quality control, quality assurance and qualified personnel other than through quality monitoring in accordance with our agreements with the CMOs. If regulatory authorities do not approve these facilities for the manufacture of our product candidates or if they withdraw 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 ARCALYST or our current or future 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 products or product candidates, if approved, operating restrictions and criminal prosecutions, any of which could significantly and adversely affect our business and supplies of our products or product candidates.

Our product candidates may also compete with other product candidates and approved products for access to and capacity within manufacturing facilities. There are a limited number of manufacturers that operate under cGMP regulations and that might be capable of manufacturing for us. Furthermore, given the limited number of available manufacturing slots and the long lead times needed to reserve them, manufacturers require monetary commitments in connection with such reservations as well as fees for changes or cancellations in the reserved manufacturing slots. As a result, we may wait to reserve manufacturing slots until we can be informed by data from the clinical trials of our product candidates, which may be several months from the time we request manufacturing slots. Any significant delay in the supply of clinical materials for our product candidates could considerably delay conducting our clinical trials and potential regulatory approval of our product candidates. Alternatively, we may project when we may need additional clinical material for our product candidates and reserve manufacturing time-slots "at-risk" prior to our product candidates having generated data from their then current clinical trials.

In addition, given the lead times we must provide to Regeneron with respect to the commercial supply of ARCALYST, we must place purchase orders based on projected demand, in advance of knowing the market acceptance of ARCALYST for the treatment of recurrent pericarditis. Such projections involve risks and uncertainties. For example, we may be unable to swiftly accommodate for unforeseen increases in commercial demand for ARCALYST given the lead times we must provide to Regeneron and limitations on Regeneron's manufacturing capacity for ARCALYST. These risks may result in additional costs or delays in manufacturing clinical materials for our product candidates when

and if we actually need them and commercial materials for ARCALYST and may result in having too little or too much of our product candidates or ARCALYST in inventory to meet actual demand.

Any performance failure on the part of our existing or future manufacturers could delay, as applicable, clinical development or marketing approval or commercialization efforts for ARCALYST and our product candidates, if approved. If our current CMOs cannot perform as agreed, we may be required to replace such manufacturers. Although we believe that there are several potential alternative manufacturers who could manufacture our product candidates, we may incur added costs and delays in identifying and qualifying any such replacement. In addition, we may not be able to establish new agreements on acceptable terms, if at all, with such alternative manufacturers. Further, Regeneron has an exclusive right to produce ARCALYST, subject to limited exceptions, which could impact our ability to find a replacement manufacturer for ARCALYST in a short period of time, if needed. Additionally, establishing a replacement manufacturer for ARCALYST or our product candidates, if required, is unlikely to be accomplished in a timely or cost-effective manner, 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, results of operations and financial condition. If we or our CMOs 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.

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 ARCALYST or 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 our safety procedures and the safety procedures utilized by our third-party manufacturers and suppliers for handling and disposing of these materials generally comply with the standards prescribed by these laws and regulations, we cannot guarantee that this is the case or eliminate the risk of accidental contamination or injury from these materials. In such an event, we may be held liable for any resulting damages and such liability could exceed our resources, and state or federal or other applicable authorities may curtail our use of certain materials or interrupt our business operations. Furthermore, environmental laws and regulations are complex, change frequently and have tended to become more stringent. We cannot predict the impact of such changes and cannot be certain of our future compliance. We do not currently carry biological or hazardous waste insurance coverage.

Manufacturing or supply issues could cause product shortages, disrupt or delay our clinical trials or regulatory approvals, delay or stop commercialization of our products and product candidates, and adversely affect our business.

The manufacture of ARCALYST and 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 ARCALYST or our product candidates failing to meet approved specifications, failed batches or other failures, such as defective products or manufacturing failures. We have limited experience overseeing the manufacturing processes of ARCALYST, mavrilimumab, vixarelimab and KPL-404. Due to the highly technical requirements of manufacturing ARCALYST and our product candidates and the strict quality and control specifications, we and our third-party providers may be unable to manufacture or supply ARCALYST or 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, as applicable, which may lead to lawsuits or could delay the introduction of our product candidates to the market.

The manufacture of ARCALYST and our product candidates is at high risk of product loss due to contamination, equipment malfunctions, human error or raw material variability or shortages. Deviations from

established manufacturing processes could result in reduced production yields, failed batches and other supply disruptions and increased costs. If microbial, viral or other contaminations are discovered in ARCALYST or 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. The involvement of our third-party manufacturers, including Regeneron, may exacerbate such effects, which has required and may in the future require us to reject lots for quality control purposes. See "*Risk Factors—We contract with third parties for manufacturing our commercial supply of ARCALYST and clinical supply for our product candidates and for certain research and other preclinical development and expect that we will continue to do so in the future. This reliance on third parties increases the risk that we may not have sufficient quantities of ARCALYST or our product candidates or such quantities at an acceptable cost, which could delay, prevent or impair our development or commercialization efforts*."

Many additional factors could cause production interruptions at our facilities or at the facilities of our third-party providers, as well as disruptions in travel, shipping or delivery capabilities into and within the countries in which we or our manufacturers produce ARCALYST or our product candidates or disruptions to production capabilities, including due to the impact of natural disasters, accidents, boycotts, labor disputes, political and economic instability, such as acts of terrorism or war, including the ongoing war in Ukraine, and an epidemic or pandemic or other outbreak of disease, including the COVID-19 pandemic. The occurrence of any such event could adversely affect our ability to satisfy the required supply for any of ARCALYST or our product candidates or successfully complete preclinical and clinical development, which would result in additional costs to us or impair our ability to generate revenue and would harm our business, financial condition and prospects significantly.

Supply chain issues related to important ancillary products may also adversely affect our business. For example, we contract with a network of specialty pharmacies who distribute ARCALYST as well as peripheral supplies that are required to reconstitute and self-administer ARCALYST, such as sterile water for injection, syringes and needles. A delay or shortage in the supply or the distribution of the peripheral supplies required to administer ARCALYST may impact patient access to ARCALYST and could cause us to lose potential revenue, reduce our potential profitability, and damage our reputation.

We also contract with third parties to source specialized placebo for use in our clinical trials which cannot be easily replaced as it must be nearly indistinguishable from our product candidates to ensure proper clinical trial blinding. If we encounter shortages of such placebo, our clinical trials may be substantially delayed unless and until we can source suitable replacements.

In addition, our third-party providers may fail to comply with cGMP and other stringent regulatory requirements related to the manufacturing process. See "*Risk Factors—We contract with third parties for manufacturing our commercial supply of ARCALYST and clinical supply for our product candidates and for certain research and other preclinical development and expect that we will continue to do so in the future. This reliance on third parties increases the risk that we may not have sufficient quantities of ARCALYST or our product candidates or such quantities at an acceptable cost, which could delay, prevent or impair our development or commercialization efforts*."

We and our third-party providers are required to maintain compliance with cGMP and other stringent requirements and are subject to inspections by the FDA and comparable agencies in other jurisdictions to confirm such compliance. If we or any of our third-party providers are not able to establish and maintain procedures and processes sufficient to satisfy cGMP or similar foreign standards, we could experience a delay, interruption or other issues in our manufacture, fill-finish, packaging, storage or delivery of ARCALYST or our product candidates, and any related failure of the facilities or operations of third parties to pass any regulatory agency inspection could significantly impair our ability to supply our products and product candidates. Significant noncompliance could also result in the imposition of monetary penalties or other civil or criminal sanctions and damage our reputation.

Any adverse developments affecting the operations of our third-party providers, such as any impact due to the COVID-19 pandemic including shortages or reprioritizations of raw materials, reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, could result in a shortage of commercial products or product candidates, the imposition of additional commercial product requirements by regulatory authorities, the withdrawal of our product candidates or approved products, shipment delays,

lot failures or recalls. We may also have to write off inventory and incur other charges and expenses for products that fail to meet specifications, undertake costly remediation efforts or seek more costly manufacturing alternatives. Such manufacturing issues could increase our cost of goods, cause us to lose potential revenue, reduce our potential profitability or damage our reputation.

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

The drug substance and drug product used in ARCALYST, mavrilimumab and vixarelimab are supplied to us from single-source suppliers and we obtain the drug substance and drug product used in KPL-404 from a limited number of sources. Regeneron has a contractual right to be our sole source manufacturer of ARCALYST unless they have a persistent failure to satisfy our supply needs. Our ability to continue to commercialize ARCALYST, to develop our product candidates, and to ultimately supply our commercial products in quantities sufficient to meet market demand, depends in part on our ability to obtain the drug substance and drug product for ARCALYST and these product candidates in accordance with regulatory requirements and in sufficient quantities for commercialization and clinical testing. With respect to ARCALYST, mavrilimumab and vixarelimab, we do not currently have arrangements in place for a redundant or second-source supply of any such drug substance and drug product in the event any of our current suppliers of such drug substance and drug product cease their operations or stop offering us sufficient quantities of these materials for any reason. With respect to KPL-404, while we anticipate having more than one source for drug substance and drug product, such sources are nonetheless limited and subject to similar risks as our other products and product candidates.

We are not certain that our single-source suppliers will be able to meet our demand for our products and product candidates, either because of the nature of our agreements with those suppliers, our limited experience with those suppliers or our relative importance as a customer to those suppliers. It may be difficult for us to assess their ability to timely meet our demand in the future based on past performance. While our suppliers have generally met our demand on a timely basis in the past, they may subordinate our needs in the future to their other customers.

In addition to manufacturing our products and product candidates in the quantities that we believe would be required to meet anticipated market demand, our third-party manufacturers may need to increase manufacturing capacity and, in some cases, alternative sources of commercial supply may need to be secured, which could involve significant challenges and may require additional regulatory approvals. In addition, the development of commercial-scale manufacturing capabilities may require us and our third-party manufacturers to invest substantial additional funds and hire and retain the technical personnel who have the necessary manufacturing experience. Neither we nor our third-party manufacturers may successfully complete any required increase to existing manufacturing capacity in a timely manner, or at all.

Moreover, our ability to progress our preclinical and clinical programs or successfully commercialize our products could be materially and adversely impacted if any of the third-party suppliers upon which we rely for raw materials and preclinical and clinical stage product candidate and commercial stage product supply were to experience a significant business challenge, disruption or failure due to issues such as financial difficulties or bankruptcy, issues relating to other customers such as regulatory or quality compliance issues, or other financial, legal, regulatory or reputational issues. Additionally, any damage to or destruction of our manufacturing facilities or equipment or those of our third-party manufacturers' or suppliers' facilities or equipment may significantly impair our ability to manufacture our products and product candidates on a timely basis.

In addition to the above, we have entered into, and may enter into in the future, collaboration and other agreements requiring us to provide commercial or clinical drug supply to third-party partners. A failure by our CMOs to supply sufficient quantities of drug supply may cause us to breach our contractual obligations, triggering potential penalties under our agreements, including termination of such agreements, if we fail to adequately cure such breach.

Establishing additional or replacement suppliers for the drug substance and drug product used in ARCALYST or our product candidates, if required, is unlikely to be accomplished quickly and can take several years, if at all.

Furthermore, despite our efforts, we may be unable to procure a replacement supplier or do so on commercially reasonable terms, which could have a material adverse impact upon our business. If we or our CMOs 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 and our CMOs may seek to maintain adequate inventory of the drug substance and drug product used in ARCALYST or our product candidates, any interruption or delay in the supply of components or materials, or our inability to obtain such drug substance and drug product from alternate sources of comparable quality at acceptable prices in a timely manner could impede, delay, limit or prevent our development or commercialization efforts, which could harm our business, results of operations, financial condition and prospects.

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

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

We rely on medical institutions, clinical investigators, contract laboratories and other third parties, such as CROs, to activate sites, conduct or otherwise support our preclinical studies and clinical trials for our product candidates properly and on time. We also rely on third parties to conduct other research related to our product candidates. We expect to rely heavily on these parties for such site activation, execution of and otherwise supporting clinical trials for our product candidates. While we have agreements governing their activities and we review the compliance history and performance of our CROs as well as have the ability to audit such activities, we have no direct control over their activities and have limited influence over their actual performance other than through quality monitoring in accordance with our agreements with the CROs. The third parties with whom we contract for execution of our preclinical studies and our clinical trials play a significant role in the conduct of these trials and the subsequent collection and analysis of data. 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 preclinical studies and clinical trials in accordance with applicable GLP or GCP requirements, we remain responsible for ensuring that each of our clinical trials is conducted in accordance with the applicable protocol, legal and regulatory requirements and scientific standards, and our reliance on these third parties does not and will not relieve us of our regulatory responsibilities. For any violations of laws and regulations during the conduct of our preclinical studies or clinical trials, we could be subject to warning letters or enforcement actions that may include civil penalties and criminal prosecution.

We and our CROs are required to comply with regulations, including GCPs, for conducting, monitoring, recording and reporting the results of clinical trials to ensure that the data and results are scientifically credible and accurate, and that the trial patients are adequately informed of the potential risks of participating in clinical trials and their rights are protected. If we or our CROs fail to comply with applicable GCPs, the clinical data generated in our clinical trials may be deemed unreliable, and regulatory authorities may require us to perform additional clinical trials before approving our marketing applications. In addition, our clinical trials must be conducted with product candidates produced under cGMPs or similar foreign regulations. Our failure or the failure of our CROs to comply with these regulations may require us to repeat clinical trials, which would delay the regulatory approval process and could also

subject us to enforcement action. We also are required to register certain clinical trials and post the results of completed clinical trials on a government-sponsored database within certain timeframes. Failure to do so when required can result in fines, adverse publicity and civil and criminal sanctions.

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

- have staffing difficulties;
- have disruptions to their business and operations, including as a result of the impact from an epidemic or pandemic disease outbreak, including COVID-19 (see "*Risk Factors — Risks related to product development — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact on our current or planned preclinical studies and clinical trials, which could be significant*") or as the result of war, conflict or terrorism as with the ongoing war in Ukraine (see "*Risk Factors – General Risk Factors – The ongoing war in Ukraine, and actions taken against Russia as a result of its invasion of Ukraine, has and may continue to have an adverse impact on the global economy, equity capital markets and our clinical operations*");
- fail to comply with contractual obligations;
- have difficulty controlling the performance of their subcontractors;
- experience regulatory compliance issues;
- undergo changes in priorities or become financially distressed; or
- form relationships with other entities, some of which may be our competitors.

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

Further, if our CROs, their subcontractors or the clinical trial sites fail to devote sufficient resources to the development of our product candidates, or if their performance is substandard, it may delay or compromise the prospects for approval and commercialization of our product candidates. In addition, the use of third-party service providers requires us to disclose our proprietary information to these parties, which could increase the risk that this information is misappropriated.

If the third parties conducting our preclinical studies or our clinical trials do not perform their contractual duties or obligations, experience significant business challenges, disruptions or failures, such as due to the impact of the COVID-19 pandemic, do not meet expected deadlines, terminate their agreements with us or need to be replaced, or if

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

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

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

See also, "*Risk Factors – Risks Related to Intellectual Property – If we are unable to protect the confidentiality of our trade secrets, our business and competitive position may be harmed.*"

Risks Related to Competition, Executing our Strategy 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.

We are not aware of any FDA-approved therapies for recurrent pericarditis, but are aware of two programs being developed in this indication. One is by R-Pharm International (RPH-104), which inhibits IL-1α/IL-1β-induced signaling and is in Phase 2 development and the other is an oral cannabidiol being developed by Cardiol Therapeutics in an open label Phase 2 setting. Anakinra (KINERET), marketed by Swedish Orphan Biovitrum AB, is currently approved for use in RA, CAPS and DIRA. We are not aware of any active, industry sponsored development programs using anakinra seeking a label for recurrent pericarditis. Canakinumab (ILARIS), marketed by Novartis Pharmaceuticals Corporation, is currently approved for use in CAPS, Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS), Mevalonate Kinase Deficiency (MKD) and Familial Mediterranean Fever (FMF), Still's Disease and Systemic Juvenile Idiopathic Arthritis (SJIA). We are not aware of any active, industry sponsored development programs using canakinumab seeking a label for recurrent pericarditis. Additionally, Novartis is also developing gevokizumab for use in oncologic indications. There are other therapies which modulate IL-1α in preclinical and clinical development for diseases other than recurrent pericarditis from Johnson &

Johnson and XBIOTECH USA, INC. We are not aware of any active, industry sponsored development programs for these programs seeking a label for recurrent pericarditis.

There are five other programs in clinical development in various indications that modulate GM-CSF signaling from GlaxoSmithKline plc, or GSK (otilimab), I-MAB Biopharma (plonmarlimab), Roivant Sciences Ltd. (gimsilumab and namilumab) and Humanigen, Inc. (lenzilumab). All of these competitive programs target the GM-CSF ligand itself versus targeting the GM-CSF receptor like mavrilimumab.

There are various programs in clinical development antagonizing the CD40 / CD154 costimulatory pathway, however KPL-404 is the only program designed to be administered subcutaneously. Certain programs are designed for intravenous administration only. Astellas Pharma Inc. is developing bleselumab (anti-CD40); Horizon Therapeutics plc is developing the Tn3 fusion protein, dazodalibep (anti-CD40L); Biogen, Inc. and UCB S.A. are developing dapirolizumab pegol (anti-CD40L); and, Eledon Pharmaceuticals, Inc. are developing AT-1501 (anti-CD40L). Certain other programs pose the potential for subcutaneous administration. Novartis A.G. is developing CFZ-533, or iscalimab (anti-CD40), Sanofi S.A./ImmuNext Inc. are developing SAR441344 (anti-CD40L), Bristol Myers-Squibb is developing BMS-986325 (anti-CD40); and Abbvie, Inc., Boehringer Ingelheim International GmbH are developing ravagalimab (anti-CD40), and H. Lundbeck A/S is developing Lu AG22515 (anti-CD40L).

Further, the results of clinical trials for our product candidates may produce negative, inconclusive or uncompetitive results compared to those produced by any of these or other companies in the indications we are studying, which may result in us deciding, or regulatory authorities requiring us, to conduct additional clinical trials or modify or cease development programs for our product candidates. We may also determine that the potential product and commercial profile of any of our product candidates may not ultimately be commercially successful or even if they have the potential to ultimately be successful, we may not have sufficient recourses, which in either case could lead us to discontinue its development in certain indications, or we may determine to not support further development of any of our product candidates at any time for any reason.

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

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

We may not be successful in executing our growth strategy to identify, discover, develop, in-license or acquire additional product candidates or technologies, and our growth strategy may not deliver the anticipated results or we may refine or otherwise alter our growth strategy. We may seek to acquire businesses or undertake business combinations, collaborations or other strategic transactions which may not be successful or on favorable terms, if at all, and we may not realize the intended benefits of such transactions.

We have acquired or in-licensed our existing product candidates, and as part of our strategy we plan to identify new product candidates or technologies that we believe are complementary to our existing product candidates. We may

do this through our internal discovery program, or by acquiring the rights to product candidates and technologies through a variety of transaction types, including in-licensing, strategic transactions, mergers or acquisitions. If we are unable to identify, discover, develop, in-license or otherwise acquire and integrate product candidates, or their related companies, in accordance with this strategy, our ability to pursue this component of our growth strategy would be limited and we may need to refine or otherwise alter this strategy. We cannot be certain that we will be successful in such efforts, and even if we are successful in such efforts, we cannot be certain that such discovery or transaction will be on favorable terms, or that, following any such discovery or transaction, we will be able to realize the intended benefits of it.

Research programs and business development efforts to identify new product candidates and technologies require substantial technical, financial and human resources. We may focus our efforts and resources on potential product candidates, technologies or businesses that ultimately prove to be unsuccessful. In-licensing and acquisitions of product candidates, technology or businesses often require significant payments and expenses and consume additional resources. We will need to continue to devote a substantial amount of time and personnel to research, develop and commercialize any such in-licensed or acquired product candidate or technology, or integrate any new business, and we may decide to reprioritize our efforts even after having expended resources on a particular prospect. Our research programs and business development efforts, including businesses or technology acquisitions, collaborations or licensing attempts, may fail to yield additional complementary or successful product candidates for clinical development and commercialization or successful business combinations for a number of reasons, including, but not limited to, the following:

- our research or business development methodology or search criteria and process may be unsuccessful in identifying potential product candidates or businesses with a high probability of success for development progression;
- we may not be able or willing to assemble sufficient resources or expertise to in-license, acquire or discover additional product candidates or acquire businesses or undertake business combinations, collaborations, or other strategic transactions;
- we may not be able to agree to acceptable terms with potential licensors or other partners or with respect to business acquisitions; and
- any product candidates or technologies to which we acquire the rights or that we discover may not allow us to leverage our expertise and our development and commercial infrastructure as currently expected.

If any of these events occurs, we may not be successful in executing our growth strategy to identify, discover, develop, in-license or acquire additional product candidates or technologies or to acquire businesses or undertake business combinations, collaborations, or other strategic transactions, or our growth strategy or strategic transactions may not deliver the anticipated results or we may refine or otherwise alter this strategy.

The consummation or performance of any acquisition, business combination, collaboration or other strategic transaction we may undertake in furtherance of our growth strategy or any refined or otherwise altered strategy, may involve additional risks, such as difficulties in assimilating different workplace cultures, retaining personnel and integrating operations, which may be geographically dispersed, increased costs, exposure to liabilities, incurrence of indebtedness, or use a substantial portion of our available cash for all or a portion of the consideration or cause dilution to our existing shareholders if we issue equity securities for all or a portion of the consideration. If any of these events occurs or we are unable to meet our strategic objectives for any such transaction, we may not be able to achieve the expected benefits from the transaction and our business may be materially harmed.

We may seek to enter into collaboration, licensing or other strategic transactions or arrangements to further develop, commercialize or otherwise attempt to realize value from one or more of our product candidates, and any such transactions or arrangements that we may enter into may not be successful or be on favorable terms, which could adversely affect our ability to develop, commercialize or attempt to realize value from our product candidates.

We have entered into and may seek to enter into collaboration, licensing or other strategic transactions or arrangements to further develop, commercialize or otherwise attempt to realize value from one or more of our product candidates instead of developing or commercializing our product candidates ourselves. For example, in February 2022, we granted Huadong exclusive rights to develop and commercialize rilonacept and mavrilimumab in the Asia Pacific region, excluding Japan. In August 2022, we entered into a license agreement with Genentech where we granted exclusive worldwide rights to develop and commercialize vixarelimab. We may seek to jointly develop, commercialize or otherwise exploit one or more of our other product candidates with a third party. To the extent that we decide to enter into such transactions or arrangements, we may face significant competition in seeking appropriate collaborators, licensees or other strategic partners. Moreover, these transactions and arrangements are complex and time consuming to negotiate, document, implement and to close or maintain. We may not be successful in our efforts to establish collaborations, licenses or other strategic transactions or arrangements should we choose to do so. The terms of any such transactions or arrangements that we may establish may have unfavorable tax consequences for our shareholders in the United States. Further, granting territory-specific rights for our products and product candidates may reduce their attractiveness for subsequent business development activity. In addition, our right to grant a sublicense of intellectual property licensed to us under certain of our current agreements requires the consent of the applicable licensor.

Any current or future collaborations, licenses or other strategic transactions or arrangements that we enter into may not be successful. The success of these potential collaboration, license arrangements and other strategic transactions or arrangements may depend heavily on the efforts and activities of our collaborators, sublicensees or other strategic partners. We have experienced collaboration failure in the past and may experience similar failures in the future. Collaborations, licenses or other strategic transactions or arrangements are subject to numerous risks, which may include risks that the collaborator, licensee or other strategic partner, as applicable:

- may have significant discretion in determining the efforts and resources that they will apply;
- may not commit sufficient resources to or otherwise not perform satisfactorily in carrying out its activities;
- may not properly prosecute, maintain or defend our intellectual property rights or may use our intellectual property or proprietary information in a way that gives rise to actual or threatened litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential liability;
- may own or co-own intellectual property covering products that results from our arrangement with them, that is not properly prepared, prosecuted, maintained or defended in a way that could impact that patentability of the intellectual property or validity for any granted patent, which could shorten the term during which we are owed royalties on such intellectual property;
- may own or co-own intellectual property covering products that results from our arrangement with them, and in such cases, we would not have the exclusive right to develop or commercialize such intellectual property, and even if we are able to license such exclusive rights, we may have to enter into a license agreement that include obligations to make milestone, royalty or other payments under such agreement;
- may delay, dispute or refuse to pay milestone and royalty payments, which may materially impact the collaboration revenue that we realize from such relationship and impact our ability to satisfy upstream payment obligations, if applicable; and
- may conduct sales and marketing activities or other operations that may not comply with applicable laws, resulting in civil or criminal proceedings.

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

We need to continue to develop our company and expand our scope of operations, and we may encounter difficulties in managing this development and expansion, which could disrupt our operations.

We expect to continue to develop our company and expand the scope of our operations. To manage our anticipated development and expansion, we must continue to implement and improve our managerial, operational and financial systems and infrastructure, expand our facilities over time and continue to recruit and train qualified personnel. Also, our executive and senior management teams have and may continue to divert a disproportionate amount of their attention away from their day-to-day activities and devote a substantial amount of time to managing these development and expansion activities. For example, in January 2021, we implemented select components of a new ERP system that will enable the organization to manage the complexity of operating a commercial organization more efficiently. As with any implementation, this new system will require specific skills and expertise to setup, maintain and utilize the system. We may not be able to develop these skills internally or in sufficient time and capacity, which could require us to expend additional resources to acquire them. Due to our limited resources, certain employees have and may continue to perform activities that are beyond their regular scope of work, and we may not be able to effectively manage the development of our company, expansion of our operations or recruit and train qualified personnel. This may result in weaknesses of our systems and infrastructure, give rise to managerial, operational and financial mistakes, loss of business opportunities, loss of employees and reduced productivity among remaining employees. The development of our company and expansion of our operations may lead to significant costs and may divert financial resources from other projects, such as the development of one or more of our product candidates. If our executive and senior management teams are unable to effectively manage our anticipated 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 as planned, including with respect to our commercialization of ARCALYST in recurrent pericarditis. Our future financial performance and our ability to commercialize our product candidates, if approved, and to compete effectively will depend, in part, on our ability to effectively manage the future development of our company and expansion of our operations.

Risks Related to Intellectual Property

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

Our commercial success depends in part on our ability to obtain and maintain proprietary or intellectual property protection in the United States and other countries for our products and product candidates, including ARCALYST, mavrilimumab, and KPL-404. 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 products and product candidates in an effort to establish intellectual property positions directed to their compositions of matter and manufacture as well as uses of these products and 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 ARCALYST, an exclusive license under the MedImmune Agreement to patent applications and patents relating to mavrilimumab, and an exclusive license under our license agreement with Beth Israel Deaconess Medical Center to patent applications and patents related to KPL-404.

Certain provisions in our intellectual property agreements may be susceptible to multiple interpretations. The resolution of any contract interpretation disagreement that may arise could affect the scope of our rights to the relevant

intellectual property or technology, or affect financial or other obligations under the relevant agreement, either of which could have a material adverse effect on our business, financial condition, results of operations and prospects.

We or our licensors have not pursued or maintained, and may not pursue or maintain in the future, patent protection for our products or product candidates in every country or territory in which we may sell our products or product candidates, if approved. In addition, we cannot be sure that any of our pending patent applications or pending trademark applications will issue or that, if issued, they will be in a form that is 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 ARCALYST, mavrilimumab, KPL-404, or any future products and product candidates. A U.S. patent covering ARCALYST as a composition of matter expired in 2020, and relevant composition of matter patents issued outside of the United States are expected to expire by 2023, not including any patent term extensions. A U.S. patent covering methods of using ARCALYST in the treatment of recurrent pericarditis issued in June 2021 and has a statutory term that expires in 2038, not including any patent term adjustment. The composition of matter patents for mavrilimumab generally have statutory expiration dates in 2027, not including any extensions or adjustments. The issued composition of matter patents for KPL-404 have statutory expiration dates in 2036, not including any extensions. The issued composition of matter patents licensed from Beth Israel Deaconess Medical Center related to KPL-404 have statutory expiration dates in 2032, not including any patent term extensions or adjustments. 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. For example, the applicable regulatory exclusivity period is often triggered by the date a product candidate obtains regulatory approval, and we cannot predict with any certainty whether and if so, when, the applicable product would receive regulatory approval in any given jurisdiction. Furthermore, the type, scope and duration of such exclusivities will vary on a country-by-country basis depending on the jurisdiction in which a product candidate is approved and the particular regulatory exclusivity for which the product is eligible as of the time of approval in such jurisdiction.

Patents may be eligible for limited patent term extension in the United States under the Drug Price Competition and Patent Term Restoration Act of 1984, referred to as the Hatch-Waxman Act. Similar patent extensions exist in the EU and Japan, subject to the applicable laws in those jurisdictions. In most countries, including the United States, the patent term is 20 years from the earliest filing date of a non-provisional patent application. In the United States, a patent's term may be lengthened by patent term adjustment, which compensates a patentee for administrative delays by the USPTO, in examining and granting a patent, or may be shortened if a patent is terminally disclaimed over an earlier expiring patent. In certain countries, the term of a patent that covers a drug product may also be eligible for patent term extension when regulatory approval is granted, provided the legal requirements are met. We may not receive an extension if we fail to apply within applicable deadlines or fail or are unable 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 ARCALYST for the treatment of CAPS in 2008, 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 ARCALYST for CAPS, or any other indication for which the FDA may grant approval in the future, is unavailable. Further, while patent term extension was awarded for relevant patents in certain European countries following the EMA's approval of ARCALYST for the

treatment of CAPS, in 2012 the marketing authorization for CAPs was withdrawn. Patent term extensions may no longer be in effect or available, subject to the applicable laws in those countries as well as other factors, such as whether a marketing approval for ARCALYST is reissued and whether such reissuance is prior to the expiration of the patent's natural 20-year patent term. Moreover, the length of the extension could be less than we request. In addition, the laws of other countries may not protect our rights to the same extent as the laws of the United States. 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 or product candidate. For example, the patents in the United States covering ARCALYST as a composition of matter have expired, and patents covering ARCALYST as a composition of matter in Europe have a term that expires in 2023, not including any patent term extensions, and the patents covering mavrilimumab as a composition of matter have a term that expires in 2027 in the United States, not including any patent term adjustments or patent term extensions, and in 2027 in Europe, not including any patent term extensions. We may not receive any patent term extension for patents covering mavrilimumab as a composition of matter if such patent in an applicable jurisdiction expires before mavrilimumab would be eligible to receive regulatory approval in such jurisdiction. 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. The expiration date of regulatory exclusivity is determined on a country-by country-basis if the applicable product is approved in such country and if any applicable regulatory exclusivity applies and is granted. The actual expiration date of any such regulatory exclusivity, however, is subject to significant uncertainty. For instance, the applicable regulatory exclusivity period is often triggered by the date a product candidate obtains regulatory approval, and we cannot predict with any certainty whether and if so, when, the applicable product would receive regulatory approval in any given jurisdiction. Furthermore, the type, scope and duration of such exclusivities will vary on a country-by-country basis depending on the jurisdictions in which a product candidate is approved and the particular regulatory exclusivity for which the product is eligible as of the time of approval.

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

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

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

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

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

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

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

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

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

We entered into agreements to acquire the rights to develop and commercialize ARCALYST and our product candidates, mavrilimumab 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 ARCALYST. In December 2017, we entered into the MedImmune Agreement to obtain exclusive worldwide rights to research, develop, manufacture, market and sell mavrilimumab and any other products covered by the licensed patent rights. In connection with our acquisition of Primatope in March 2019, we acquired an exclusive world-wide license with Beth Israel Deaconess Medical Center for certain patent applications and patents related to KPL-404. Each of these agreements requires us to use commercially reasonable efforts to develop and commercialize the related product candidates, make timely milestone and other payments, provide certain information regarding our activities with respect to such product candidates and indemnify the other party with respect to our development and commercialization activities under the terms of the agreements. These agreements and any future such agreements that we enter into impose a variety of obligations and related consequences.

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

- our diligence obligations with respect to the use of the licensed technology in relation to our development and commercialization of our product candidates, and what activities satisfy those diligence obligations;
- the scope of rights granted under the agreement and other interpretation-related issues;
- our obligations to make milestone, royalty or other payments under those agreements;
- whether and the extent to which our technology and processes infringe on intellectual property of the licensor that is not subject to the agreement;
- our right to sublicense patent and other rights to third parties;
- the ownership of inventions and know-how resulting from the joint creation or use of intellectual property by our licensors and us and our partners;
- our right to transfer or assign the license; and

- the effects of termination.

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

If we fail to meet our obligations under these agreements in a material respect, the respective licensor/seller would have the right to terminate the respective agreement and upon the effective date of such termination, have the right to re-obtain the related technology as well as aspects of any intellectual property controlled by us and developed during the period the agreement was in force that relate to the applicable technology. This means that the licensor/seller to each of these agreements could effectively take control of the development and commercialization of our product candidates after an uncured, material breach of the agreement by us. This would also be the case if we voluntarily elected to terminate the relevant agreement, which we have the right to do under each of these agreements. While we would expect to exercise our rights and remedies available to us in the event we fail to meet our obligations under these agreements in any material respect, including seeking to cure any breach by us, and otherwise seek to preserve our rights under the technology licensed to or acquired by us, we may not be able to do so in a timely manner, at an acceptable cost or at all. Any uncured, material breach under the license could result in our loss of exclusive rights and may lead to a complete termination of our product development and any commercialization efforts for each of our product candidates. Termination of one of these agreements for any reason, and the related discontinuation of the development or commercialization of a product candidate could impair our ability to raise additional capital, generate revenue and may significantly harm our business, financial condition and prospects.

The FDA approved ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years of age and older in March 2021. Upon receipt of this approval, we assumed the sales and distribution of ARCALYST for the previously approved indications in the United States. We evenly split profits on ARCALYST sales with Regeneron. Regeneron retains worldwide rights to develop and commercialize ARCALYST for local administration to the eye and ear and oncology. Additionally, Regeneron retains the right to develop and commercialize ARCALYST for all applications in the Middle East and North Africa. The development of ARCALYST in other fields could increase the possibility of identification of adverse safety results that impact the commercialization of ARCALYST for the treatment of recurrent pericarditis.

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 Regeneron Agreement, Regeneron has a right of first negotiation over the assignment or sale of our rights to any product we develop under the Regeneron Agreement to third parties and we must obtain Regeneron's prior consent to assign or sublicense our rights under such agreement to a third party. 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.

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 products and product candidates, if approved, 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 products, 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 products and product candidates and technology, including contested proceedings before the USPTO. Our competitors or other third parties may assert infringement claims against us, alleging that our products are covered by their patents.

Given the vast number of patents in our field of technology, we cannot be certain that we do not infringe existing patents or that we will not infringe patents that may be granted in the future. Many companies have filed, and continue to file, patent applications related to immunomodulation. Some of these patent applications have already been allowed or issued, and others may issue in the future. For example, we are aware of third-party patents that contain claims potentially relevant to mavrilimumab and KPL-404. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to mavrilimumab and KPL-404 would be found to infringe any valid claim of these patents. While we may decide to initiate proceedings to challenge the validity of these or other patents in the future, we may be unsuccessful, and courts or patent offices in the United States and abroad could uphold the validity of any such patent. If we were to challenge the validity of any issued U.S. patent in court, we would need to overcome a statutory presumption of validity that attaches to every U.S. patent. This means that in order to prevail, we would have to present clear and convincing evidence as to the invalidity of the patent's claims. In order to avoid infringing these or any other third-party patents, we may find it necessary or prudent to obtain licenses to such patents from such third-party intellectual property holders. However, we may be unable to secure such licenses or otherwise acquire or in-license any compositions, methods of use processes or other intellectual property rights from third parties that we identify as necessary for our current or future product candidates. The licensing or acquisition of third-party intellectual property rights is a competitive area, and several more established companies may also pursue strategies to license or acquire third-party intellectual property rights that we may consider attractive or necessary. These established companies may have a competitive advantage over us due to their size, capital resources and greater clinical development and commercialization capabilities. In addition, companies that perceive us to be a competitor may be unwilling to assign or license rights to us. We also may be unable to license or acquire third-party intellectual property rights on terms that would allow us to make an appropriate return on our investment or at all. If we are unable to successfully obtain rights to required third-party intellectual property or maintain the existing intellectual property rights we have, we may have to cease development of the relevant program or product candidate, which could have a material adverse effect on our business, financial condition, results of operations and prospects.

Since our product candidates are being developed for use in fields that are competitive and of strong interest to pharmaceutical and biotechnology companies, we will likely seek to file additional patent applications and may have additional patents granted in the future, based on our future research and development efforts. Furthermore, because patent applications can take many years to issue and may be confidential for 18 months or more after filing, and because pending patent claims can be revised before issuance, there may be applications of third-parties 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.

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 infringement, 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, non-enablement, or foreign equivalents thereof. 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 or unenforceable.

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

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

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

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

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

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

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

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

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

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

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

As is the case with other biopharmaceutical companies, our success is heavily dependent on intellectual property, particularly patents. Obtaining and enforcing patents in the biopharmaceutical industry involve both technological and legal complexity and is therefore costly, time consuming and inherently uncertain. Patent reform legislation in the United States and other countries, including the Leahy-Smith America Invents Act, or Leahy-Smith Act, signed into law on September 16, 2011, contribute to those uncertainties and costs. The Leahy-Smith Act includes a number of significant changes to U.S. patent law. These include provisions that have affected the way patent applications are prosecuted, and have redefined prior art and provided 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 in which, assuming that other requirements of patentability are met, the first inventor to file a patent application will be entitled to the patent regardless of whether a third party was first to invent the claimed invention. A third party that has filed a patent application in the USPTO after March 2013 but before us could therefore be awarded a patent covering an invention of ours even if we had made the invention before it was made by such third party. This requires us to be cognizant of the time from invention to filing of a patent application. Furthermore, our ability to obtain and maintain valid and enforceable patents depends on whether the differences between our technology and the prior art allow our technology to be patentable over the prior art. Since patent applications in the United States and most other countries are confidential for a period of time after filing or until issuance, we cannot be certain that we were the first to either (1) file any patent application related to our product candidates or (2) invent any of the inventions claimed in our patents or patent applications. Even where we have a valid and enforceable patent, we may not be able to exclude others from practicing the claimed invention where the other party can show that they used the invention in commerce before our filing date or the other party benefits from a compulsory license.

Among some of the other changes introduced by the Leahy-Smith Act are changes that (i) affect the way patent applications are prosecuted, (ii) redefine prior art, and (iii) provide more efficient and cost-effective avenues for competitors to challenge the validity of patents. These include 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. Accordingly, a third party may attempt to use the USPTO procedures to invalidate our patent claims that would not have been invalidated if first challenged by the third party as a defendant in a district court action. Thus, the Leahy-Smith Act and its implementation increase the uncertainties and costs surrounding the prosecution of our or our future licensors' patent applications and the enforcement or defense of our issued patents, all of which could have a material adverse effect on our business, financial condition, results of operations and prospects.

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

In Europe, beginning no earlier than October 1, 2022, European patent applications will have the option, upon grant of a patent, of becoming a "Unitary Patent," which will be subject to the jurisdiction of a new patent adjudication body, the Unitary Patent Court (UPC). The UPC is intended to provide an alternative mechanism to disputing patent

rights in the courts of individual European countries. The UPC will be a significant change in European patent practice. Since the UPC is a new court, there are no existing precedents and the rulings of the UPC will be unpredictable at the outset, thus increasing the uncertainty of any litigation involving a Unitary Patent.

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

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

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

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

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

See also "*Risk Factors – Risks Related to Manufacturing and Our Reliance on Third Parties – 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*."

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

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

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

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

General Risk Factors

The ongoing war in Ukraine, and actions taken against Russia as a result of its invasion of Ukraine, has and may continue to have an adverse impact on the global economy, equity capital markets and our clinical operations.

In February 2022, Russia invaded Ukraine to the condemnation of the international community. Although the length, impact and outcome of the ongoing military conflict in Ukraine is highly unpredictable, this conflict could lead to significant market and other disruptions, including significant volatility in commodity prices and supply of energy resources, instability in financial markets, supply chain interruptions, political and social instability, changes in consumer or purchaser preferences as well as increases in cyberattacks and espionage. In response to Russia’s invasion and perpetration of war crimes, western nations, including the United States and the EU, instated severe economic sanctions against Russia, including the suspension of Russian banks from the global SWIFT system, travel bans, personal sanctions against the Russian elite and boycotts of Russian goods and commodities. Private organizations responded similarly, with many companies choosing to suspend their Russian operations or end them entirely.

The situation is rapidly evolving, and the United States, the EU, the UK and other countries may implement additional sanctions, export controls or other measures against Russia, Belarus and other countries, regions, officials, individuals or industries in the respective territories. Such sanctions and other measures, as well as the existing and potential further responses from Russia or other countries to such sanctions, tensions and military actions, could adversely affect the global economy and financial markets and could adversely affect our business, operations and financial position. The prices of certain commodities have and will likely continue to increase in the wake of the conflict.

U.S. capital markets have seen significant volatility as a result of the conflict. The war may also pose risks to global supply chains, further threatening the global and U.S. economy. The price of our shares and the shares of other biopharmaceutical companies may fluctuate or otherwise be impacted, especially if the war or governmental responses thereto result in a prolonged economic downturn. As a result of such disruptions, we may be unable to raise additional capital when needed or on acceptable terms, if at all. These risks may also be compounded with the effects of the COVID-19 pandemic, and measures taken in response to the pandemic, on U.S. and global capital markets. See "*Risk Factors – General Risk Factors – The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position.*"

The conflict in Ukraine has and may continue to adversely affect our clinical development efforts. We conduct clinical trials at sites across the globe, including in Eastern Europe. In the aftermath of the invasion, we terminated planned clinical trial operations in Ukraine and Russia. While we anticipate being able to secure alternative clinical trial sites in Western Europe, there is no guarantee that such sites will be able to provide a sufficient number of patients to satisfy our clinical need and at a reasonable cost. Further, should the conflict escalate beyond Ukraine, we may need to take further actions. In such an instance, we may be unable to secure alternative clinical sites when needed or on acceptable terms, if at all. The foregoing may cause significant delays or disruptions to our clinical development efforts, which could have a material impact on our business, operations and financial position.

Furthermore, due to the political uncertainty involving Russia and Ukraine, there is also an increased likelihood that we or our CROs or other third parties with whom we conduct business or otherwise engage, may also be subject to retaliatory cyberattacks perpetrated by Russia or others at its direction in response to economic sanctions and other actions taken against Russia as a result of its invasion of Ukraine. See "*Risk Factors – General Risk Factors – Our information technology systems, or those of our third party CMOs, CROs or other contractors, consultants and service providers, may fail or suffer cyberattacks or security breaches, which could result in a material disruption of our or such third-party's business or operations and our development programs for our product candidates or loss of other assets, including funds*."

The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position.

The COVID-19 pandemic, and measures taken in response to the pandemic, could cause significant disruption in our business and operations and could cause significant disruption in the business and operations of our manufacturers, CROs upon whom we rely to conduct our clinical trials, clinical trial sites, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities.

The federal and state governments in the United States and the governments of other countries around the globe implemented various measures in response to the COVID-19 pandemic, including significant restrictions on businesses as well as travel into and within the countries in which our manufacturers produce our product candidates or where we conduct our clinical trials or otherwise conduct business or engage with other third parties. While many such restrictive measures have since been loosened or repealed following the widespread distribution of COVID-19 vaccines, the loosening of such restrictions have been and may in the future be subject to abrupt reversal in the presence of new variants of COVID-19, including the Omicron variants, which may lengthen or exacerbate the pandemic's effect on our business, financial condition and results of operations. In addition, government restrictions and policies remain unpredictable and varied across jurisdictions, introducing significant uncertainty and compliance difficulty.

If the COVID-19 pandemic is prolonged, including through the emergence of new variants of the virus, and measures undertaken in response to the pandemic are reinstated, we may experience and our manufacturers, CROs or other third parties with whom we conduct business or otherwise engage, may experience or continue to experience

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

In response to the COVID-19 pandemic and measures introduced by state and federal governments in the United States, we implemented workplace protocols at our facilities. We have required all employees entering our workplaces in the United States to be fully vaccinated against COVID-19, subject to a reasonable accommodation process. We have also established additional safety measures at our facilities, including providing and requiring the use of personal protective equipment, testing prior to returning to the office after an exposure and/or onset of symptoms, enforcing occupancy limits, and implementing enhanced contact tracing tools. These safety measures do not, however, guarantee that COVID-19 will not spread amongst our employees through workplace contact, and the sickness of employees may have a significant adverse impact on our business. We continue to monitor the developments, restrictions and requirements in jurisdictions where we have offices, and plan to update the protocols for our offices as applicable.

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

Further, the COVID-19 pandemic may impact our ability to successfully commercialize ARCALYST and our current and future product candidates, if approved. The COVID-19 pandemic and measures taken in response to the pandemic, including business and travel restrictions and social distancing to halt the spread of the pandemic, has had an impact on businesses, healthcare systems, regulatory authorities and other organizations and conferences. These measures may result in limitations on certain aspects of our commercialization strategy, including our specialty cardiology salesforce having limited, or zero, access to prescriber offices and other high-volume accounts in person, which if prolonged, may impede the effective commercialization of ARCALYST and result in lower than anticipated future revenue.

As a result of the COVID-19 pandemic, existing and any new third party CMOs or suppliers may be unable to produce or supply our current or future products and product candidates or obtain certain raw or fill-finish materials, including vials and stoppers, needed to produce or supply such products and product candidates. Such parties may also experience delays, restrictions or limitations in the production, delivery or release of our products and product candidates or the raw or fill-finish materials needed to produce them, including due to disruptions at their respective facilities, staffing shortages, production slowdowns, stoppages or reprioritizations, including as a result of reprioritization by third parties or the U.S. government for any products or potential products related to the treatment or prevention of COVID-19, or interruptions in global shipping. Any failure to source sufficient quantities of our products and product candidates could prevent us from successfully commercializing our approved products and/or delay or force us to cancel our clinical activity.

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

The United Kingdom's withdrawal from the European Union may have a negative effect on global economic conditions, financial markets and our business.

Following a national referendum and enactment of legislation by the government of the UK, the UK formally withdrew from the EU and ratified a trade and cooperation agreement governing its future relationship with the EU, referred to as Brexit. The agreement, which was being applied provisionally from January 1, 2021 and entered into force on May 1, 2021, addresses trade, economic arrangements, law enforcement, judicial cooperation and a governance framework including procedures for dispute resolution, among other things. Because the agreement merely sets forth a framework in many respects and will require complex additional bilateral negotiations between the UK and the EU as both parties continue to work on the rules for implementation, significant political and economic uncertainty remains about how the precise terms of the relationship between the parties will differ from the terms before withdrawal.

Since January 1, 2021, however, the UK has operated under a distinct regulatory regime to the EU. EU pharmaceutical laws only apply in respect of the UK to Northern Ireland (as set out in the Protocol on Ireland/Northern Ireland). EU laws which have been transposed into UK law through secondary legislation continue to be applicable as "retained EU law". The UK government has proposed to repeal the majority of this retained EU law by the end of 2023, which may lead to further regulatory uncertainty and could result in cost increases for our business. While the UK has indicated a general intention that new laws regarding the development, manufacture and commercialization of medicinal products in the UK will align closely with EU law, there are limited detailed proposals for future regulation of medicinal products. The trade and cooperation agreement includes specific provisions concerning medicinal products, which include the mutual recognition of GMP, inspections of manufacturing facilities for medicinal products and GMP documents issued (such mutual recognition can be rejected by either party in certain circumstances), but does not foresee wholesale mutual recognition of UK and EU pharmaceutical regulations. For example, it is not clear to what extent the UK will adopt legislation aligned with, or similar to, the EU CTR which became applicable on January 31, 2022 and which significantly reforms the assessment and supervision processes for clinical trials throughout the EU. On January 17, 2022, the Medicines and Healthcare products Regulatory Agency, or MHRA, launched an eight-week consultation on reframing the UK legislation for clinical trials. The consultation closed on March 14, 2022 and aims to streamline clinical trials approvals, enable innovation, enhance clinical trials transparency, enable greater risk proportionality, and promote patient and public involvement in clinical trials. The outcome of the consultation will be closely watched and will determine whether the UK chooses to align with the regulation or diverge from it to maintain regulatory flexibility. A decision by the UK not to closely align its regulations with the new approach that will be adopted in the EU may have an effect on the cost of conducting clinical trials in the UK as opposed to other countries and/or make it harder to seek a marketing authorization in the EU for our product candidates on the basis of clinical trials conducted in the UK.

Therefore, there remains political and economic uncertainty regarding to what extent the regulation of medicinal products will differ between the UK and the EU in the future. Any divergences will increase the cost and complexity of running our business, including with respect to the conduct of clinical trials. Brexit also materially impacted the regulatory regime with respect to the approval of our product candidates. Great Britain is no longer covered by the EU's procedures for the grant of marketing authorizations (Northern Ireland is covered by the centralized authorization procedure and can be covered under the decentralized or mutual recognition procedures). As of January 1, 2021, all existing centralized marketing authorizations were automatically converted into UK marketing authorizations effective in Great Britain and issued with a United Kingdom marketing authorization number on January 1, 2021 (unless marketing authorization holders opted out of this scheme). A separate marketing authorization is now required to market drugs in Great Britain. It is currently unclear whether the regulator in the UK, the MHRA, is sufficiently prepared to handle the increased volume of marketing authorization applications that it is likely to receive. The UK's withdrawal from the EU and the associated uncertainty has had and may continue to have a significant adverse effect on global economic conditions and the stability of global financial markets, and could significantly reduce global market liquidity and restrict the ability of key market participants to operate in certain financial markets. Asset valuations, currency exchange rates and credit ratings may be especially subject to increased market volatility. The UK has also experienced significant political instability in 2022, which has seen three different Prime Ministers hold office. Any of these factors could have a significant adverse effect on our business, financial condition, results of operations and prospects.

Further, the UK's withdrawal from the EU has resulted in the relocation of the EMA from the UK to the Netherlands. This relocation has caused, and may continue to cause, disruption in the administrative and medical scientific

links between the EMA and the MHRA, including delays in granting clinical trial authorization or marketing authorization, disruption of importation and export of active substance and other components of new drug formulations, and disruption of the supply chain for clinical trial product and final authorized formulations. The cumulative effects of the disruption to the regulatory framework may add considerably to the development lead time to marketing authorization and commercialization of products in the EU and/or the UK.

Such a situation could hinder our ability to conduct current and planned clinical trials and commercialize our products and product candidates, if approved, including ARCALYST, adversely affecting our business, financial condition and results of operations. Additionally, political instability in the EU as a result of Brexit may result in a material negative effect on credit markets and foreign direct investments in the EU and UK.

These developments, or the perception that any related developments could occur, have had and may continue to have a material adverse effect on global economic conditions and the financial markets, and may significantly reduce global market liquidity, restrict the ability of key market participants to operate in certain financial markets or restrict our access to capital. Any of these factors could have a material adverse effect on our business, financial condition and results of operations and reduce the price of our shares.

If we fail to comply with reporting and payment obligations under the Medicaid Drug Rebate Program or other governmental pricing programs, we could be subject to additional reimbursement requirements, penalties, sanctions and fines, which could have a material adverse effect on our business, financial condition, results of operations and growth prospects.

We participate in governmental programs that impose extensive drug price reporting and payment obligations on pharmaceutical manufacturers. Medicaid is a joint federal and state program that is administered by the states for low-income and disabled beneficiaries. Under the Medicaid Drug Rebate Program (the "MDRP"), as a condition of federal funds being made available for our covered outpatient drugs under Medicaid and certain drugs or biologicals under Medicare Part B, we pay a rebate to state Medicaid programs for each unit of our covered outpatient drugs dispensed to a Medicaid beneficiary and paid for by the state Medicaid program. Medicaid rebates are based on pricing data that we report on a monthly and quarterly basis to CMS, the federal agency that administers the MDRP and Medicare programs. For the MDRP, these data include the Average Manufacturer Price ("AMP") for each drug and, in the case of innovator products, best price. If we become aware that our MDRP price reporting submission for a prior period was incorrect or has changed as a result of recalculation of the pricing data, we must resubmit the corrected data for up to three years after those data originally were due. If we fail to provide information timely or are found to have knowingly submitted false information to the government, we may be subject to civil monetary penalties and other sanctions, including termination from the MDRP, in which case payment would not be available for our covered outpatient drugs under Medicaid or, if applicable, Medicare Part B.

Federal law requires that any company that participates in the MDRP also participate in the 340B program in order for federal funds to be available for the manufacturer's drugs under Medicaid and Medicare Part B. The 340B program is administered by HRSA and requires us, as a participating manufacturer, to agree to charge statutorily defined covered entities no more than the 340B "ceiling price" for our covered drugs used in an outpatient setting. These 340B covered entities include a variety of community health clinics and other entities that receive health services grants from the Public Health Service, as well as hospitals that serve a disproportionate share of low-income patients. A drug that is designated for a rare disease or condition by the Secretary of Health and Human Services is not subject to the 340B ceiling price requirement with regard to the following types of covered entities: rural referral centers, sole community hospitals, critical access hospitals, and free-standing cancer hospitals. The 340B ceiling price is calculated using a statutory formula, which is based on the AMP and rebate amount for the covered outpatient drug as calculated under the MDRP. In general, products subject to Medicaid price reporting and rebate liability are also subject to the 340B ceiling price calculation and discount requirement. We must report 340B ceiling prices to HRSA on a quarterly basis, and HRSA publishes them to 340B covered entities. HRSA has finalized regulations regarding the calculation of the 340B ceiling price and the imposition of civil monetary penalties on manufacturers that knowingly and intentionally overcharge covered entities for 340B-eligible drugs. HRSA has also finalized an administrative dispute resolution process through which 340B covered entities may pursue claims against participating manufacturers for overcharges, and through which manufacturers may pursue claims against 340B covered entities for engaging in unlawful diversion or

duplicate discounting of 340B drugs. In addition, legislation may be introduced that, if passed, would further expand the 340B program, such as adding further covered entities or requiring participating manufacturers to agree to provide 340B discounted pricing on drugs used in an inpatient setting.

In order to be eligible to have drug products paid for with federal funds under Medicaid and Medicare Part B and purchased by certain federal agencies and grantees, we must also participate in the VA/FSS pricing program. Under the VA/FSS program, we must report the Non-FAMP for our covered drugs to the VA and charge certain federal agencies no more than the Federal Ceiling Price, which is calculated based on Non-FAMP using a statutory formula. These four agencies are the VA, the U.S. Department of Defense, the U.S. Coast Guard, and the U.S. Public Health Service (including the Indian Health Service). We must also pay rebates on products purchased by military personnel and dependents through the TRICARE retail pharmacy program. If we fail to provide timely information or are found to have knowingly submitted false information, we may be subject to civil monetary penalties.

Individual states continue to consider and have enacted legislation to limit the growth of healthcare costs, including the cost of prescription drugs and combination products. A number of states have either implemented or are considering implementation of drug price transparency legislation that may prevent or limit our ability to take price increases at certain rates or frequencies. Requirements under such laws include advance notice of planned price increases, reporting price increase amounts and factors considered in taking such increases, wholesale acquisition cost information disclosure to prescribers, purchasers, and state agencies, and new product notice and reporting. Such legislation could limit the price or payment for certain drugs, and states may impose civil monetary penalties or pursue other enforcement mechanisms against manufacturers who fail to comply with drug price transparency requirements. If we are found to have violated state law requirements, we may become subject to penalties or other enforcement mechanisms, which could have a material adverse effect on our business.

Pricing and rebate calculations vary across products and programs, are complex, and are often subject to interpretation by us, governmental or regulatory agencies, and the courts, which can change and evolve over time. Such pricing calculations and reporting, along with any necessary restatements and recalculations, could increase our costs for complying with the laws and regulations governing the MDRP and other governmental programs, and under the MDRP could result in an overage or undercharge in Medicaid rebate liability for past quarters. Price recalculations under the MDRP also may affect the ceiling price at which we are required to offer products under the 340B program. Civil monetary penalties can be applied if we are found to have knowingly submitted any false price or product information to the government, if we fail to submit the required price data on a timely basis, or if we are found to have charged 340B covered entities more than the statutorily mandated ceiling price. CMS could also terminate our Medicaid drug rebate agreement, in which case federal payments may not be available under Medicaid or Medicare Part B, if applicable, for our covered outpatient drugs. Pursuant to the Inflation Reduction Act of 2022 (the “IRA”), the AMP figures we report will also be used to compute rebates under Medicare Part D triggered by price increases that outpace inflation. We cannot assure you that our submissions will not be found to be incomplete or incorrect.

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

In the United States, EU and other jurisdictions, there have been and we expect there will continue to be a number of legislative and regulatory initiatives and proposed changes to the healthcare system that could affect our operations. For example, in the United States, the Affordable Care Act (the “ACA”) substantially changed the way healthcare is financed by both governmental and private insurers, and significantly impacts the U.S. pharmaceutical industry. The ACA, among other things, subjected biologic products to potential competition by lower-cost biosimilars, addressed a new methodology by which rebates owed by manufacturers under the MDRP are calculated for drugs and biologics that are inhaled, infused, instilled, implanted or injected, increased the minimum Medicaid rebates owed by manufacturers under the MDRP, extended manufacturer Medicaid rebate obligations to utilization by individuals enrolled in Medicaid managed care organizations, established annual fees and taxes on manufacturers of certain branded prescription drugs and biologics, and established a new Medicare Part D coverage gap discount program.

Since its enactment, there have been judicial, executive and Congressional challenges to certain aspects of the ACA. In June 2021, the U.S. Supreme Court dismissed the most recent judicial challenge to the ACA brought by several

states without specifically ruling on the constitutionality of the ACA. In addition, other legislative changes have been proposed and adopted in the United States since the ACA was enacted. The Budget Control Act of 2011, among other things, led to reductions of Medicare payments to providers, which will remain in effect through 2031 unless additional Congressional action is taken. More recently, in March 2021, President Biden signed into law the American Rescue Plan Act of 2021, which eliminates the statutory cap on the Medicaid drug rebate, currently set at 100% of a drug's AMP, beginning January 1, 2024.

Most significantly, in August 2022, President Biden signed the IRA into law. This statute marks the most significant action by Congress with respect to the pharmaceutical industry since adoption of the ACA in 2010. Among other things, the IRA requires manufacturers of certain drugs to engage in price negotiations with Medicare (beginning in 2026), with prices that can be negotiated subject to a cap; imposes rebates under Medicare Part B and Medicare Part D to penalize price increases that outpace inflation (first due in 2023); and replaces the Part D coverage gap discount program with a new discounting program (beginning in 2025). The IRA permits the Secretary of the Department of Health and Human Services (HHS) to implement many of these provisions through guidance, as opposed to regulation, for the initial years. For that and other reasons, it is currently unclear how the IRA will be effectuated, and while the impact of the IRA on our business and the pharmaceutical industry cannot yet be fully determined, it is likely to be significant.

Additionally, there has been increasing legislative and enforcement interest in the United States with respect to specialty drug pricing practices. Specifically, there have been several recent Congressional inquiries and proposed and enacted federal and state legislation designed to, among other things, bring more transparency to drug and biologic pricing, reduce the cost of prescription drugs and biologics under Medicare, review the relationship between pricing and manufacturer patient programs and reform government program reimbursement methodologies for drugs and biologics. For example, in May 2019, CMS published a final rule to allow Medicare Advantage Plans the option to use step therapy for Part B drugs beginning January 1, 2020. In addition, in March 2021, the American Rescue Plan Act of 2021 was signed into law, which eliminates the statutory Medicaid drug rebate cap, currently set at 100% of a drug's average manufacturer price, beginning January 1, 2024. Most recently, in August 2022, the IRA was signed into law, which implemented a number of significant changes, as described above. 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 products and product candidates, if approved, or additional pricing pressures.

Individual states and municipalities 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, marketing cost disclosure and other transparency measures, and, in some cases, measures 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 products and product candidates, if approved, or put pressure on our product pricing.

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

In markets outside of the United States and the EU, reimbursement and healthcare payment systems vary significantly by country, and many countries have instituted price ceilings on specific products and therapies. In the UK, the recent political instability has resulted in Rishi Sunak being appointed as the UK's third Prime Minister of 2022. Given the new UK government is in its infancy, there remains significant uncertainty as to whether reforms are likely to be made to healthcare legislation in the UK.

We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative action, either in the United States, the EU or elsewhere. For example, such actions may result in changes to governmental policies and regulations that affect our operations and business, including our clinical trials, regulatory approval, pharmaceutical pricing and reimbursement. If we or any third party we may engage are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we or such third party are not able to maintain regulatory compliance, our product candidates may lose any regulatory approval that may have been obtained and we may not achieve or sustain corporate profitability.

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

Our results of operations could be adversely affected by general conditions in the global economy and in the global financial markets. For example, the COVID-19 pandemic, the ongoing war in Ukraine and increases in inflation and interest rates have impacted and continue to impact the global economy, causing or contributing to global supply chain issues, price fluctuations and other significant economic effects. See "*Risk Factors — General Risk Factors — The COVID-19 pandemic, and measures taken in response to the pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position"* and "*Risk Factors — General Risk Factors — The ongoing war in Ukraine, and actions taken against Russia as a result of its invasion of Ukraine, has and may continue to have an adverse impact on the global economy, equity capital markets and our clinical operations."* In addition, global credit and financial markets have recently experienced volatility and disruptions, including severely diminished liquidity and credit availability, rising interest rates, declines in consumer confidence, declines in economic growth, increase in unemployment rates and uncertainty about economic stability.

These disruptions could adversely affect our ability to manufacture, market and sell our commercialized products, including ARCALYST, and satisfy the required supply for any of our product candidates or successfully complete preclinical and clinical development of our product candidates, which could require us to incur additional costs, and impair our ability to obtain regulatory approval of our product candidates and generate revenue. Doing business internationally involves a number of other risks, including but not limited to:

- multiple, conflicting and changing laws and regulations such as privacy regulations, tax laws, employment laws, regulatory requirements, permits and export and import restrictions;
- failure by us to obtain and maintain regulatory approvals for the use of our products in various countries;
- additional potentially relevant third-party patent rights;
- complexities and difficulties in obtaining protection and enforcing our intellectual property;
- difficulties in staffing and managing operations outside of the United States;
- complexities associated with managing multiple payor reimbursement regimes, government payors or patient self-pay systems;
- limits in our ability to penetrate international markets;

- financial risks, such as longer payment cycles, difficulty collecting accounts receivable, the impact of local and regional financial crises on demand and payment for our products and exposure to foreign currency exchange rate fluctuations;

- natural disasters, political and economic instability such as war, including the ongoing war in Ukraine, 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 clinical activities, sales and other functions that may fall within the purview of the U.S. Foreign Corrupt Practices Act, its books and records provisions or its antibribery 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 information technology systems, or those of our third party CMOs, CROs or other contractors, consultants and service providers, may fail or suffer cyberattacks or security breaches, which could result in a material disruption of our or such third-party's business or operations and our development programs for our product candidates or loss of other assets, including funds.

Despite the implementation of security measures, our information technology systems and those of our third-party CMOs, CROs and other contractors, consultants and service providers as well as employees that are working outside of our facilities are vulnerable to attack, damage or interruption from viruses and malware (e.g., ransomware), malicious code, theft, natural disasters, terrorism, war, telecommunication and electrical failures, hacking, cyberattacks, phishing attacks and other social engineering schemes, employee misuse, human error, fraud, denial or degradation of service attacks, sophisticated nation-state and nation-state-supported actors or unauthorized access or use by persons inside our organization, or persons with access to systems inside our organization. Attacks upon information technology systems are increasing in their frequency, levels of persistence, sophistication and intensity, and are being conducted by sophisticated and organized groups and individuals with a wide range of motives and expertise. As a result of the COVID-19 pandemic, we may face increased cybersecurity risks due to our reliance on internet technology and the number of our employees who are working remotely, which may create additional opportunities for cybercriminals to exploit vulnerabilities. Employees may also fail to comply with our cybersecurity protocols, exposing us to vulnerabilities despite our safeguards. Furthermore, because the techniques used to obtain unauthorized access to, or to sabotage, systems change frequently and often are not recognized until launched against a target, we may be unable to anticipate these techniques or implement adequate preventative measures. In addition, a breach at a CMO, CRO, contractor, consultant, service provider or other third party with which we engage may increase our exposure by allowing criminals to exploit our relationship with such persons. Such security breaches may remain undetected for an extended period. Even if identified, we may be unable to adequately investigate or remediate incidents or breaches due to attackers increasingly using tools and techniques that are designed to circumvent controls, to avoid detection, and to remove or obfuscate forensic evidence.

We and certain of our service providers are from time to time subject to cyberattacks and security incidents. While we do not believe that we have experienced any significant system failure, accident or security breach to date, if such an event were to occur and cause interruptions in our business and operations or those of our third party CMOs, CROs and other contractors, consultants and service providers as well as employees that are working outside of our facilities, the costs associated with the investigation, remediation and potential notification of a breach to counter-parties and data subjects could be material. A breach could result in a material disruption of our or such third-party's business or operations and our development programs of our product candidates' or loss of other assets, including funds. For example, the loss of clinical trial data for our product candidates could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any disruption or security breach results in a loss of or damage to our data or applications or other data or applications relating to our technology or

product candidates, or inappropriate disclosure or theft of confidential or proprietary information, the further development of our product candidates could be delayed. Although we maintain cybersecurity insurance coverage, it may not be adequate to cover all liabilities that we may incur from cyber-attacks or security breaches and is subject to deductibles and coverage limitations.

Actual or perceived failures to comply with applicable data protection, privacy and security laws, regulations, standards and other requirements could adversely affect our business, results of operations, and financial condition.

We are or in the future may be subject to data privacy and protection laws, regulations, policies and contractual obligations that govern the collection, transmission, storage, processing, and use of personal information or personal data. The regulatory framework for data privacy and security worldwide is continuously evolving and developing and, as a result, interpretation and implementation standards and enforcement practices are likely to remain uncertain for the foreseeable future. This evolution may affect our ability to operate in certain jurisdictions or to collect, store, transfer use and share personal information, necessitate the acceptance of more onerous obligations in our contracts, result in liability or impose additional costs on us. The cost of compliance with these laws, regulations and standards is high and is likely to increase in the future. Any failure or perceived failure by us to comply with federal, state or foreign laws or regulations, our internal policies and procedures or our contracts governing our processing of personal information could result in negative publicity, government investigations and enforcement actions, claims by third parties and damage to our reputation, any of which could have a material adverse effect on our business, results of operation, and financial condition.

For example, 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. We do not believe that we are currently acting as a covered entity or business associate under HIPAA and thus are not subject to its requirements or penalties. However, any person may be prosecuted under HIPAA's criminal provisions either directly or under aiding-and-abetting or conspiracy principles. Consequently, depending on the facts and circumstances, we could face substantial criminal penalties if we knowingly receive individually identifiable health information from a HIPAA-covered healthcare provider or research institution that has not satisfied HIPAA's requirements for disclosure of individually identifiable health information. In addition, we may maintain sensitive personally identifiable information, including health information, that we receive throughout the clinical trial process, in the course of our research collaborations, and directly from individuals (or their healthcare providers) who enroll in our patient support program. As such, we may be subject to state laws requiring notification of affected individuals and state regulatory authorities in the event of a breach of personal information, which is a broader class of information than the health information protected by HIPAA.

In addition, certain state laws govern the privacy and security of health information in certain circumstances, some of which are more stringent than HIPAA and many of which differ from each other in significant ways and may not have the same effect, thus complicating compliance efforts. Failure to comply with these laws, where applicable, can result in the imposition of significant civil and/or criminal penalties and private litigation. Certain states have also adopted comparable privacy and security laws and regulations governing the privacy, processing and protection of personal information. For example, California enacted the California Consumer Privacy Act, or the CCPA, on June 28, 2018, which went into effect on January 1, 2020. The CCPA gives California residents expanded rights to access, correct, and delete their personal information, opt out of certain personal information sharing and receive detailed information about how their personal information is used. The CCPA provides for civil penalties for violations, as well as a private right of action for data breaches that is expected to increase data breach litigation. Further, the California Privacy Rights Act, or CPRA, recently passed in California. The CPRA significantly amends the CCPA and will impose additional data protection obligations on covered businesses, including additional consumer rights processes, limitations on data uses, new audit requirements for higher risk data, and opt outs for certain uses of sensitive data. It will also create a new California data protection agency authorized to issue substantive regulations and could result in increased privacy and information security enforcement. Virginia, Colorado, Connecticut and Utah have also enacted similar legislation to California increasing complexity of compliance with privacy laws. The majority of the provisions for these laws will go into effect in 2023, and additional compliance investment and potential business process changes may be required.

Furthermore, the Federal Trade Commission, or FTC, and many state Attorneys General continue to enforce federal and state consumer protection laws against companies for online collection, use, dissemination and security practices that appear to be unfair or deceptive. For example, according to the FTC, failing to take appropriate steps to keep consumers' personal information secure can constitute unfair acts or practices in or affecting commerce in violation of Section 5(a) of the Federal Trade Commission Act. The FTC expects a company's data security measures to be reasonable and appropriate in light of the sensitivity and volume of consumer information it holds, the size and complexity of its business, and the cost of available tools to improve security and reduce vulnerabilities.

Our clinical trial programs outside the United States may implicate international data protection laws, including the General Data Protection Regulation 2016/679, or GDPR, and legislation of EU member states and European Economic Area, or EEA, countries implementing it. The GDPR went into effect in May 2018 and imposes strict requirements for processing the personal data of individuals within the EEA. Companies that must comply with the GDPR face increased compliance obligations and risk, including more robust regulatory enforcement of data protection requirements and potential fines for noncompliance of up to €20 million or 4% of the annual global revenues of the noncompliant company, whichever is greater. Among other requirements, the GDPR regulates transfers of personal data subject to the GDPR to third countries that have not been found to provide adequate protection to such personal data, including the United States, and the efficacy and longevity of current transfer mechanisms between the EU and the United States remains uncertain. For example, in 2016, the EU and United States agreed to a transfer framework for data transferred from the EU to the United States called the Privacy Shield, but in July 2020 the Court of Justice of the EU, or the CJEU, limited how organizations could lawfully transfer personal data from the EEA to the United States by invalidating the Privacy Shield for purposes of international transfers and imposing further restrictions on use of the standard contractual clauses, or SCCs. In March 2022, the EU and United States announced a new regulatory regime intended to replace the invalidated regulations; however, this new EU-US Data Privacy Framework has not been implemented beyond an executive order signed by President Biden on October 7, 2022 on Enhancing Safeguards for United States Signals Intelligence Activities. And while the CJEU upheld the adequacy of the SCCs, it made clear that reliance on them alone may not necessarily be sufficient in all circumstances. Use of the SCCs must now be assessed on a case-by-case basis taking into account the legal regime applicable in the destination country, in particular applicable surveillance laws and rights of individuals and additional measures and/or contractual provisions may need to be put in place, however, the nature of these additional measures is currently uncertain. The CJEU went on to state that if a competent supervisory authority believes that the SCCs cannot be complied with in the destination country and the required level of protection cannot be secured by other means, such supervisory authority is under an obligation to suspend or prohibit that transfer. The European Commission issued revised SCCs on June 4, 2021 to account for the decision of the CJEU and recommendations made by the European Data Protection Board. The revised SCCs must be used for relevant new data transfers from September 27, 2021; existing standard contractual clauses arrangements must be migrated to the revised clauses by December 27, 2022. There is some uncertainty around whether the revised clauses can be used for all types of data transfers, particularly whether they can be relied on for data transfers to non-EEA entities subject to the GDPR. The revised SCCs apply only to the transfer of personal data outside of the EEA and not the UK. The UK's Information Commissioner's Office has published new data transfer standard contracts for transfers from the UK under the UK GDPR. This new documentation will be mandatory for relevant data transfers from September 21, 2022; existing standard contractual clauses arrangements must be migrated to the new documentation by March 21, 2024. As supervisory authorities issue further guidance on personal data export mechanisms, including circumstances where the SCCs cannot be used, and/or start taking enforcement action, we could suffer additional costs, complaints and/or regulatory investigations or fines, and/or if we are otherwise unable to transfer personal data between and among countries and regions in which we operate or intend to operate, it could affect the manner in which we provide our services, the geographical location or segregation of our relevant systems and operations, and could adversely affect our financial results.

Further, following the withdrawal of the UK from the EU on January 31, 2020, and the expiration of the transition period, from January 1, 2021, companies have had to comply with the GDPR and also the UK GDPR, which, together with the amended UK Data Protection Act 2018, retains the GDPR in UK national law. The UK GDPR mirrors the fines under the GDPR, e.g., fines up to the greater of €20 million (£17.5 million) or 4% of global turnover. The relationship between the UK and the EU in relation to certain aspects of data protection law remains unclear, and it is unclear how UK data protection laws and regulations will develop in the medium to longer term, and how data transfers to and from the UK will be regulated in the medium to longer term. The European Commission has adopted an adequacy

decision in favor of the UK, enabling data transfers from EU member states to the United Kingdom without additional safeguards. However, the UK adequacy decision will automatically expire in June 2025 unless the European Commission re-assesses and renews or extends that decision and remains under review by the Commission during this period. In September 2021, the UK government launched a consultation on its proposals for wide-ranging reform of UK data protection laws following Brexit. There is a risk that any material changes which are made to the UK data protection regime could result in the European Commission reviewing the UK adequacy decision, and the UK losing its adequacy decision if the European Commission deems the UK to no longer provide adequate protection for personal data.

The EU has also proposed a Regulation on Privacy and Electronic Communications, or ePrivacy Regulation, which, if adopted, would impose new obligations on the use of personal data in the context of electronic communications, particularly with respect to online tracking technologies and direct marketing. Additionally, the EU adopted the EU Clinical Trials Regulation, which came into effect on January 31, 2022. This regulation imposes new obligations on the use of data generated from clinical trials and enables European patients to have the opportunity to access information about clinical trials. Failure or perceived failure to comply with the GDPR, the UK GDPR, the ePrivacy Regulation, the EU Clinical Trials Regulations, and other countries' privacy or data security-related laws, rules or regulations could result in significant regulatory penalties and fines, affect our compliance with contracts entered into with our partners and collaborators, and could have an adverse effect on our reputation, business and financial condition.

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

Although we work to comply with applicable laws, regulations and standards, our contractual obligations and other legal obligations, these requirements are evolving and may be modified, interpreted and applied in an inconsistent manner from one jurisdiction to another, and may conflict with one another or other legal obligations with which we must comply. If we or third-party CMOs, CROs or other contractors or consultants fail to comply with applicable regulatory requirements, we could be subject to a range of regulatory actions that could affect our or our contractors' ability to develop and commercialize our product candidates and could harm or prevent sales of any affected products that we are able to commercialize, or could substantially increase the costs and expenses of developing, commercializing and marketing our products. Any threatened or actual government enforcement action could also generate adverse publicity and require that we devote substantial resources that could otherwise be used in other aspects of our business.

Securities class action and derivative lawsuits and other legal proceedings are often brought against companies, which could result in substantial costs and divert management's attention.

Securities class action and derivative lawsuits and other legal proceedings are often brought against companies following a decline in the market price of their securities. There can be significant fluctuations in market price for the securities of biopharmaceutical companies, such as us. As a result, we may be more susceptible to these types of lawsuits and legal proceedings than other companies with more stable security prices. In connection with any litigation or other legal proceedings, we could incur substantial costs, and such costs and any related settlements or judgments may not be covered by insurance. We could also suffer an adverse impact to our reputation and a diversion of management's attention and resources, which could have a material adverse effect on our business.

Although we maintain director and officer liability insurance coverage, it may not be adequate to cover all liabilities that we may incur and is subject to deductibles and coverage limitations. Insurance coverage is increasingly expensive. We may not be able to maintain insurance coverage at a reasonable cost or in an amount adequate to satisfy any liability that may arise. If we are unable to obtain insurance at acceptable cost or otherwise protect against potential class action and derivative lawsuits and other legal proceedings or claims often brought against companies following a

decline in the market price of their securities, we will be exposed to significant liabilities, which may materially and adversely affect our business and financial position.

We and our employees and third parties with whom we contract are increasingly utilizing social media tools as a means of communication both internally and externally, and noncompliance with applicable requirements, policies or contracts due to social media use or negative posts or comments could have an adverse effect on our business.

Despite our efforts to monitor evolving social media communication guidelines and comply with applicable rules, there is risk that the use of social media by us or our employees to communicate about our products, product candidates or business may cause us to be found in violation of applicable requirements. In addition, our employees or third parties with whom we contract, such as our CROs or CMOs, may knowingly or inadvertently make use of social media in ways that may not comply with our social media policy or other legal or contractual requirements, which may give rise to liability, lead to the loss of trade secrets or other intellectual property or result in public exposure of personal information of our employees, clinical trial patients, customers and others or information regarding our product candidates or clinical trials. Clinical trial patients may also knowingly or inadvertently make use of social media in ways that may not align with our communication strategies, including with respect to any adverse events they may experience, which may give rise to liability and regulatory risk. Furthermore, negative posts or comments about us or our products or product candidates in social media could seriously damage our reputation, brand image and goodwill. Any of these events could have a material adverse effect on our business, prospects, operating results and financial condition and could adversely affect the price of our Class A common shares.

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

We are highly dependent on the research and development, clinical, medical, regulatory, manufacturing, commercial and business development expertise of members of our executive and senior management teams, as well as the other members of our management, scientific and clinical teams. Although we have entered into employment agreements with our executive officers and certain members of senior management, each of them or we may terminate their employment with us at any time. We do not maintain “key person” insurance for any of our executives, senior management 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, regulatory, manufacturing and sales and marketing personnel is also critical to our success. The failure to recruit, or the loss of the services of our executive officers, senior management or other key employees could impede the achievement of our research, development and commercialization objectives, including with respect to our sales, marketing and distribution capabilities, infrastructure and organization to commercialize products for which we have obtained marketing approval and maintain proper regulatory oversight functions, any of which would seriously harm our ability to successfully implement our business strategy. Furthermore, replacing executive officers, senior management and key employees may be difficult and may take an extended period of time because of the limited number of individuals in our industry with the breadth of skills and experience required to successfully develop, gain regulatory approval of and commercialize products. Changes in our senior management may be disruptive to our business, and, if we are unable to manage an orderly transition of responsibilities, our business may be adversely affected. Competition to hire from this limited pool is intense, and we may be unable to hire, train, retain or motivate these key personnel on acceptable terms given the competition among numerous pharmaceutical and biotechnology companies for similar personnel. We also experience competition for the hiring of scientific and clinical personnel from universities and research institutions. Failure to succeed in clinical trials may make it more challenging to recruit and retain qualified scientific and clinical 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.

Our employees, principal investigators, CROs, consultants and other third-party service providers may engage in misconduct or other improper activities, including noncompliance with regulatory standards and requirements and insider trading.

We are exposed to the risk that our employees, principal investigators, CROs, consultants and other third-party service providers may engage in fraudulent conduct or other illegal activity. Misconduct by these parties could include intentional, reckless or negligent conduct or disclosure of unauthorized activities to us that violate the regulations of the FDA and other regulatory authorities, including those laws requiring the reporting of true, complete and accurate information to such authorities; healthcare fraud and abuse laws and regulations in the United States and abroad; or laws that require the reporting of financial information or data accurately.

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

It is not always possible to identify and deter misconduct by employees and other third parties. The precautions we take to detect and prevent this activity may not be effective in controlling unknown or unmanaged risks or losses or in protecting us from governmental investigations or other actions or lawsuits stemming from a failure to comply with these laws or regulations. Additionally, we are subject to the risk that a person could allege such fraud or other misconduct, even if none occurred. If any such actions are instituted against us, and we are not successful in defending ourselves or asserting our rights, those actions could have a significant impact on our business, including the imposition of civil, criminal and administrative penalties, damages, monetary fines, possible exclusion from participation in Medicare, Medicaid and other federal healthcare programs, contractual damages, reputational harm, diminished profits and future earnings and curtailment of our operations, any of which could adversely affect our ability to operate our business and our results of operations.

The increasing focus on environmental sustainability and social initiatives could increase our costs, harm our reputation and adversely impact our financial results.

There has been increasing public focus by investors, patients, environmental activists, the media and governmental and nongovernmental organizations on a variety of environmental, social and other sustainability matters. We may experience pressure to make commitments relating to sustainability matters that affect us, including the design and implementation of specific risk mitigation strategic initiatives relating to sustainability. If we are not effective in addressing environmental, social and other sustainability matters affecting our business, or setting and meeting relevant sustainability goals, our reputation and financial results may suffer. In addition, even if we are effective at addressing such concerns, we may experience increased costs as a result of executing upon our sustainability goals that may not be offset by any benefit to our reputation, which could have an adverse impact on our business and financial condition.

In addition, this emphasis on environmental, social and other sustainability matters has resulted and may result in the adoption of new laws and regulations, including new reporting requirements. New reporting requirements may be particularly difficult or expensive to comply with and, if we fail to comply, we may be required to issue financial restatements, suffer harm to our reputation or otherwise have our business be adversely impacted.

Risks Related to Ownership of Our Common Shares

The concentration of ownership of our Class B common shares, which are held primarily by our executive officers and certain other members of our senior management, and the conversion rights of the holders of our Class A1 common shares, which shares are held primarily by entities affiliated with certain of our directors, and Class B1 common shares, all of which shares are held by entities affiliated with certain of our directors means that such persons are, and such entities may in the future be, able to influence certain matters submitted to our shareholders for approval, which may have an adverse effect on the price of our Class A common shares and may result in our Class A common shares being undervalued.

Each Class A common share is entitled to one vote per Class A common share and each Class B common share is entitled to ten votes per Class B common share. Our Class A1 common shares and Class B1 common shares have no voting rights. As a result, all matters submitted to our shareholders are decided by the vote of holders of our Class A common shares and Class B common shares. As a result of the multi-class voting structure of our common shares, our executive officers and certain other members of our senior management collectively control a substantial amount of the voting power of our common shares and therefore are able to control the outcome of certain matters submitted to our shareholders for approval. As of September 30, 2022, the holders of Class A common shares accounted for approximately 66% of our aggregate voting power and the holders of Class B common shares accounted for approximately 34% of our aggregate voting power. Our executive officers and certain other members of our senior management hold Class A common shares and Class B common shares representing approximately 30% of our aggregate voting power as of September 30, 2022 and may have the ability to influence the outcome of certain matters submitted to our shareholders for approval.

However, this percentage may change depending on any conversion of our Class B common shares, Class A1 common shares or Class B1 common shares. Each holder of our Class B common shares has the ability to convert any portion of its Class B common shares into Class B1 common shares or Class A common shares at any time with advance notice to us. Each holder of our Class B1 common shares has the ability to convert any portion of its Class B1 common shares into Class A common shares or Class B common shares at any time with advance notice to us, and each holder of our Class A1 common shares has the ability to convert any portion of its Class A1 common shares into Class A common shares at any time with advance notice to us. Our Class A1 common shares and Class B1 common shares cannot be converted if, immediately prior to or following such conversion, the holder and its affiliates beneficially own, or would beneficially own, more than 4.99% of our issued and outstanding Class A common shares unless such holder provides us with 61-days' prior notice that it intends to increase, decrease or waive such threshold upon conversion. For example, as of September 30, 2022, entities affiliated with certain members of our directors could convert their Class A1 common shares and Class B1 common shares upon 61-days' prior written notice into Class A common shares and Class B common shares, respectively, which in the aggregate would result in such entities holding approximately 78% of our aggregate voting power and having the ability to control the outcome of certain matters submitted to our shareholders for approval. Due to these conversion rights, holders of our Class A1 common shares and our Class B1 common shares could, at any time with appropriate advance notice to us, significantly increase their voting control of us, which could result in their ability to significantly influence or control matters submitted to our shareholders for approval and significantly decrease the voting power of our currently outstanding Class A common shares.

These conversion rights as well as concentrated control that limit certain shareholders' ability to influence corporate matters may have an adverse effect on the price of our Class A common shares. Holders of our Class B common shares collectively control our management and affairs and are able to influence or control the outcome of certain matters submitted to our shareholders for approval, including the election of directors. Due to the conversion rights of the holders of our Class A1 and B1 common shares, entities affiliated with certain of our directors could significantly increase their voting control of us. This concentration of control might adversely affect certain corporate actions that some of our shareholders may view as beneficial, for example, by:

- delaying, deferring or preventing a change of control of us;
- impeding a merger, consolidation, takeover or other business combination involving us; or

- discouraging a potential acquirer from making a tender offer or otherwise attempting to obtain control of us.

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

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

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

- general economic, industry and market conditions, including as a result of the COVID-19 pandemic and measures taken in response to the pandemic, global political turmoil, including the war in Ukraine, and rising inflation rates;

- changes in voting control of, or sales of our shares by, our executive officers and certain other members of our senior management or entities affiliated with certain of our directors that hold our shares; and

- the other factors described in this "Risk Factors" section.

Additionally, the trading prices of biopharmaceutical companies have been highly volatile as a result of the COVID-19 pandemic. The COVID-19 outbreak continues to rapidly evolve, including through the emergence of new variants of the virus. The extent to which the outbreak may impact our business in the future, including our commercialization of ARCALYST, our preclinical studies and clinical trials, results of operations and financial condition will depend on future developments, which are highly uncertain and cannot be predicted with confidence. See "*Risk Factors—General Risk Factors – The COVID-19 pandemic, and measures taken in response to the pandemic or the easing of such measures, could have an adverse impact that is significant on our business and operations as well as the business or operations of our manufacturers, CROs and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities, and has impacted and could continue to impact the global economy, which may have a material adverse effect on our business, operations and financial position.*"

The ongoing war in Ukraine has also introduced significant market volatility and economic instability, which may materially impact our business, operations and financial position. See "*Risk Factors—General Risk Factors – The ongoing war in Ukraine, and actions taken against Russia as a result of its invasion of Ukraine, has and may continue to have an adverse impact on the global economy, equity capital markets and our clinical operations."*

Additionally, our business and share price may be affected by adverse global economic and political conditions as well as the state of the financial markets, particularly as the United States and other countries balance concerns around debt, inflation and interest rates. There can be no assurance that global economic conditions and financial markets will stabilize in the near term and that we will not experience any adverse effects that may be material to our consolidated cash flows, results of operations, financial position or our ability to access capital.

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

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

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

A significant number of our Class A common shares are issuable upon conversion of our Class B, Class A1, and Class B1 common shares, subject to certain limitations on conversion. As of September 30, 2022, approximately 1.9 million Class A common shares directly held by our executive officers and directors, inclusive of Class A common shares issuable upon conversion of our Class B, Class A1, and Class B1 common shares, were eligible for resale in the public market to the extent permitted by the provisions of Rule 144 promulgated under the Securities Act of 1933, as amended, or the Securities Act, and such rule, Rule 144. In addition, as of September 30, 2022, there were approximately 10.1 million Class A common shares subject to outstanding share options and RSUs under our equity

incentive plans that may become eligible for sale in the public market to the extent permitted by the provisions of applicable vesting schedules and Rule 144 and Rule 701 under the Securities Act.

Over a majority of our common shares are held by our executive officers and other members of our senior management team, together with entities affiliated with certain of our directors. As of September 30, 2022, on an as-converted to Class A common shares basis, these shareholders collectively held approximately 33.7 million of our Class A common shares. If any of these shareholders sell, convert or transfer, or indicate an intention to sell, convert or transfer, a substantial amount of their common shares (after certain restrictions on conversion or resale lapse), the market price of our Class A common shares could decline.

Pursuant to our amended and restated investor rights agreement, or our investors rights agreement, certain shareholders are entitled to certain registration rights with respect our Class A common shares, including Class A common shares issuable upon conversions of our Class B, Class A1, and Class B1 common shares and upon the exercise of certain rights to acquire Class A common shares, or collectively registerable securities, under the Securities Act. As of September 30, 2022, on an as-converted to Class A common shares basis, we have registered approximately 31.8 million Class A common shares held by certain holders affiliated with certain of our directors as well as certain other shareholders pursuant to our investor rights agreement, which are freely tradable without restriction under the Securities Act, to the extent permitted by Rule 144. Further, pursuant to the investors rights agreement (a) the holders affiliated with certain of our directors are entitled to certain registration rights under the Securities Act with respect to registrable securities they may own now or in the future and (b) our executive officers are also entitled to certain registration rights under the Securities Act with respect to registrable securities they may own now or in the future, including, on an as-converted to Class A common shares basis, approximately 1.7 million Class A common shares held by certain of our executive officers as of September 30, 2022. If any of these Class A common shares are sold, or if it is perceived that they will be sold, in the public market, the market price of our Class A common shares could decline.

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

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

In addition, the consummation or performance of any acquisition, business combination, collaboration or other strategic transaction we may undertake in furtherance of our growth strategy may cause dilution to our existing shareholders if we issue equity securities for all or a portion of the consideration.

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

We are currently a "smaller reporting company" as defined under the rules promulgated under the Securities Act. As a smaller reporting company, we may follow reduced disclosure requirements and do not have to make all of the disclosures that public companies that are not smaller reporting companies do.

For so long as we remain a smaller reporting company, we are permitted and intend to rely on exemptions from certain disclosure requirements that are applicable to other public companies that are not smaller reporting companies. Smaller reporting companies are able to provide simplified executive compensation disclosure and have certain other reduced disclosure obligations, including, among other things, being required to provide only two years of audited

financial statements and not being required to provide selected financial data, supplemental financial information or risk factors.

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

We incur significant costs as a result of operating as a public company, and our management is required to devote substantial time to related compliance initiatives.

As a public company, we incur significant legal, accounting and other expenses. In addition, the Sarbanes Oxley Act of 2002 and rules subsequently implemented by the SEC and The Nasdaq Global Select Market, or Nasdaq, where our Class A common shares are listed, impose various requirements on public companies, including establishment and maintenance of effective disclosure and financial controls and corporate governance practices. Our management and other personnel devote a substantial amount of time to these compliance initiatives. Moreover, these rules and regulations increase our legal and financial compliance costs and make some activities more time-consuming and costly, which increased, for example, in connection with having been a large accelerated filer in 2021 and no longer qualifying as an emerging growth company. This may require management and other personnel to divert attention from operational and other business matters to devote substantial time to public company reporting requirements.

Pursuant to Section 404, we are required to furnish a report by our management on our internal control over financial reporting. To maintain compliance with Section 404 within the prescribed period, we engage in a process to document and evaluate our internal control over financial reporting, which is both costly and challenging. In this regard, we will need to continue to dedicate internal resources, engage outside consultants and refine and revise a detailed work plan to assess and document the adequacy of internal control over financial reporting, continue steps to improve control processes as appropriate, validate through testing that controls are functioning as documented and implement a continuous reporting and improvement process for internal control over financial reporting. Despite our efforts, there is a risk that we will not be able to conclude that our internal control over financial reporting is effective as required by Section 404. This could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statements.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

We may become subject to unanticipated tax liabilities.

Although we are incorporated under the laws of Bermuda, we may become subject to income, withholding or other taxes in certain other jurisdictions by reason of our activities and operations, including the movement of assets to and between one or more foreign subsidiaries. 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-Bermudian tax liability could materially adversely affect our results of operations.

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

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

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

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

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

Our tax position could be adversely impacted by changes in tax rates, tax laws, tax practice, tax treaties or tax regulations or changes in the interpretation thereof by the tax authorities in Europe (including the United Kingdom), the United States, Bermuda, and other jurisdictions, as well as being affected by certain changes currently proposed by the Organization for Economic Cooperation 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 were to arise, it could adversely impact our tax position and our effective tax rate. For example, the UK government announced significant tax cuts, only to see such proposed tax cuts swiftly reversed following a change in Prime Minister. Given the new administration in the UK is in its infancy, there remains significant uncertainty as to any other tax policies and strategies which this or any future administration may adopt.

Failure to manage the risks associated with such changes, or misinterpretation of the laws providing such changes, could result in costly audits, interest, penalties, and reputational damage, which could adversely affect our business, results of operations and our financial condition. Our actual effective tax rate may vary from our expectation and that variance may be material. A number of factors may increase our future effective tax rates, including:

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

Pursuant to the Bermuda Economic Substance Act 2018 (as amended) and related Economic Substance Regulations (collectively, "ES Laws"), certain entities in Bermuda engaged in "relevant activities" are required to maintain appropriate physical presence in Bermuda and to satisfy economic substance requirements. The list of "relevant activities" includes carrying on as a business in any one or more of the following categories: banking, insurance, fund

management, financing and leasing, headquarters, shipping, distribution and service center, intellectual property and holding entities. Under the ES Laws, any relevant entity carrying on a relevant activity must satisfy economic substance requirements locally or face financial penalties, restriction or regulation of its business activities or may be struck off as a registered entity from the Bermuda Register of Companies. Because we are not engaged in any "relevant activities", we believe that we are not obliged to meet the economic substance requirements. We will continue to monitor our status with respect to the ES Laws and whether further action may be required in the future by the Company to comply with the ES Laws.

Governmental agencies may enact significant changes to the taxation of business entities including, among others, an increase in the corporate income tax rate, the imposition of minimum taxes or surtaxes on certain types of income, significant changes to the taxation of income derived from international operations, and an addition of further limitations on the deductibility of business interest. While certain draft legislation has been publicly released, the likelihood of these changes being enacted or implemented is unclear. We are unable to predict whether such changes will occur. If such changes are enacted or implemented, we are unable to predict the ultimate impact on our business and therefore there can be no assurance our business will not be adversely affected.

While we believe we are not a passive foreign investment company ("PFIC") for U.S. federal income tax purposes for the year ended December 31, 2021, and do not believe we will be a PFIC for U.S. federal income tax purposes for the year ending December 31, 2022, if we were to be classified a PFIC, this could result in adverse U.S. federal income tax consequences to U.S. Holders.

We completed an analysis of the Company's and its subsidiaries sources of income and character of their assets for U.S. federal income tax purposes and determined that neither the Company nor any of its subsidiaries would be classified a PFIC for the taxable year ended December 31, 2021. We plan to perform an analysis to determine whether the Company or its subsidiaries are expected to be treated as PFICs for the taxable year ending December 31, 2022, and do not believe that the Company or its subsidiaries will be treated as a PFIC for the taxable year ending December 31, 2022. However, there can be no guarantee that the Company, or its subsidiaries, will not be treated as a PFIC for any taxable period. A non-U.S. company will generally be considered as a PFIC for any taxable year if (i) at least 75% of its gross income is passive (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, or our subsidiaries, are classified as a PFIC in any year with respect to which a U.S. Holder (as defined below) owns our Class A common shares, we will continue to be treated as a PFIC with respect to such U.S. Holder in all succeeding years during which the U.S. Holder owns the Class A common shares, regardless of whether we continue to meet the PFIC test described above, unless we cease to be a PFIC and the U.S. Holder made a "qualified electing fund" election or "mark-to-market" election for (a) the first taxable year the U.S. Holder was treated as owning our shares while we were a PFIC or (b) for the taxable year in which we were a PFIC and the U.S. Holder made a "deemed sale" election or was qualified to and made a "deemed dividend" election. A "U.S. Holder" is a beneficial owner of our Class A common shares that, for U.S. federal income tax purposes, is or is treated as any of the following:

- an individual who is a citizen or resident of the United States;
- a corporation created or organized under the laws of the United States, any state thereof, or the District of Columbia;
- an estate, the income of which is subject to U.S. federal income tax regardless of its source; or
- a trust that (i) is subject to the supervision of a U.S. court and all substantial decisions of which are subject to the control of one or more "United States persons" (within the meaning of Section 7701(a)(30) of the U.S. Internal Revenue Code of 1986, as amended (the "Code")), or (ii) has a valid election in effect to be treated as a United States person for U.S. federal income tax purposes.

If we, or our subsidiaries, are classified as a PFIC for any taxable year during which a U.S. Holder holds our Class A common shares, certain adverse U.S. federal income tax consequences could apply to such U.S. Holder,

including (i) the treatment as ordinary income of any gain realized on a disposition of our shares and distributions on our shares not being qualified dividend income, (ii) the application of a deferred interest charge on the tax on such gain and distributions, and (iii) the obligation to comply with certain reporting requirements.

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

We believe we will likely be classified as a "controlled foreign corporation" (as such term is defined in the Code) for the taxable year ended December 31, 2022. Even if we were not classified as a controlled foreign corporation, certain of our non-U.S. subsidiaries could be treated as controlled foreign corporations because our group includes one or more U.S. subsidiaries. If a U.S. Holder is treated as owning (directly, indirectly or constructively) at least 10% of the value or voting power of our shares, such U.S. Holder may be treated as a "United States shareholder" (as such term is defined in the Code) with respect to us (if we are classified as a controlled foreign corporation) and each controlled foreign corporation in our group (if any). A United States shareholder of a controlled foreign corporation may be required to annually report and include in its U.S. taxable income its pro rata share of "Subpart F income," "global intangible low-taxed income," or GILTI, and investments in U.S. property by such controlled foreign corporation, regardless of whether such corporation makes any distributions. An individual that is a United States shareholder with respect to a controlled foreign corporation generally would not be allowed certain tax deductions or foreign tax credits that would be allowed to a United States shareholder that is a U.S. corporation. Failure to comply with these reporting obligations or income inclusions may subject such shareholder to significant monetary penalties and may prevent the statute of limitations with respect to such shareholder's U.S. federal income tax return for the year for which reporting was due from starting. We cannot provide any assurances that we will assist investors in determining whether such investor is treated as a United States shareholder with respect to us or any of our non-U.S. subsidiaries. Further, we cannot provide any assurances that we will furnish to any United States shareholders information that may be necessary to comply with the reporting and tax paying obligations discussed above. U.S. Holders should consult their tax advisors regarding the potential application of these rules to any investment in our Class A common shares.

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

None.

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	
10.1†+	License Agreement, dated August 2, 2022, by and among Kiniksa Pharmaceuticals (UK), Ltd., Genentech, Inc. and F. Hoffmann-La Roche Ltd.					*
10.2+	Amendment No. 2, dated August 2, 2022, to the Asset Purchase Agreement, dated September 7, 2016, by and between the Registrant and Biogen MA Inc.					*
10.3	Fifth Amendment of Sublease, dated July 27, 2022, by and between Kiniksa Pharmaceuticals Corp. and 92 Hayden Avenue Trust					*
31.1	Rule 13a-14(a) / 15d-14(a) Certification of Chief Executive Officer					*
31.2	Rule 13a-14(a) / 15d-14(a) Certification of Chief Financial Officer					*
32.1	Section 1350 Certification of Chief Executive Officer					**
32.2	Section 1350 Certification of Chief Financial Officer					**
101.INS	XBRL Instance Document – the instance document does not appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL document					***
101.SCH	Inline XBRL Taxonomy Extension Schema Document					***
101.CAL	Inline XBRL Taxonomy Extension Calculation Linkbase Document					***
101.DEF	Inline XBRL Extension Definition Linkbase Document					***
101.LAB	Inline XBRL Taxonomy Label Linkbase Document					***
101.PRE	Inline XBRL Taxonomy Extension Presentation Linkbase Document					***
104	Cover page Interactive Data File (formatted in Inline XBRL and contained in Exhibit 101) - The cover page interactive data file does not appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL document					***

* Filed herewith
** Furnished herewith
*** Submitted electronically herewith
† Portions of the exhibit have been redacted in compliance with Regulation S-K Item 601(a)(6)
+ Portions of this exhibit have been omitted pursuant to Item 601(b)(10)(iv) of Regulation S-K

SIGNATURES

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

KINIKSA PHARMACEUTICALS, LTD.

Date: November 3, 2022

By: /s/ Mark Ragosa
Mark Ragosa
Senior Vice President and Chief Financial Officer
(Principal Financial Officer)

License Agreement

This License Agreement is entered as of the Execution Date (as defined below), with certain provisions that go into effect as of the Effective Date (as defined below),

by and between

Genentech, Inc.

with an office and place of business at 1 DNA Way, South San Francisco, California 94080, ("**Genentech**");

and

F. Hoffmann-La Roche Ltd

with an office and place of business at Grenzacherstrasse 124, 4070 Basel, Switzerland ("**Roche**," Roche and Genentech together referred to as "**GNE**")

on the one hand

and

Kiniksa Pharmaceuticals (UK), Ltd.

with an office and place of business at 23 Old Bond Street, London, UK, W1S 4PZ ("**Kiniksa**")

on the other hand.

[**] Certain information in this document has been excluded pursuant to Regulation S-K, Item 601(a)(6)

[***] Certain information in this document has been excluded pursuant to Regulation S-K, Item 601(b)(10)(iv). Such excluded information is not material and would likely cause competitive harm to the registrant if publicly disclosed.

RECITALS

WHEREAS, Kiniksa is developing vixarelimab, known as KPL-716 (the **"Lead Antibody**"), a fully human monoclonal antibody that targets the cytokine receptor subunit, oncostatin M (OSM) receptor (OSMR (as defined below)), and possesses proprietary technology and intellectual property rights relating thereto;

WHEREAS, GNE has expertise in the research, development, manufacture and commercialization of pharmaceutical products;

WHEREAS, GNE wishes to develop for commercialization the Licensed Antibodies and explore their potential applications;

WHEREAS, Kiniksa is willing to grant to GNE rights to use certain of its intellectual property rights to make, use, offer for sale, sell and import and export Licensed Antibodies and Licensed Products in the Territory for use in the Field (as such terms are respectively defined below), as contemplated herein; and

WHEREAS, GNE and Kiniksa agree that Kiniksa will perform certain activities to complete certain ongoing clinical studies, provide technology transfer, supply existing inventory and materials and manufacture certain quantities of clinical supply of Lead Antibody.

NOW, THEREFORE, in consideration of the mutual covenants and promises contained in this Agreement and other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, the Parties hereto, intending to be legally bound, do hereby agree as follows:

1. Definitions

As used in this Agreement, the following terms, whether used in the singular or plural, shall have the following meanings:

1.1 *Affiliate*

The term "**Affiliate**" shall mean, as of any point in time and so long as such relationship continues to exist, any individual, corporation, association or other business entity that directly or indirectly controls, is controlled by, or is under common control with the Party in question. As used in this definition of "Affiliate," the term "control" shall mean the direct or indirect ownership of more than fifty percent (>50%) of the stock having the right to vote for directors thereof or the ability to otherwise control the management of the corporation or other business entity whether through the ownership of voting securities, by contract, resolution, regulation or otherwise. Notwithstanding the foregoing, neither [***] will be considered an Affiliate of Kiniksa solely by reason of such control. Anything to the contrary in this paragraph notwithstanding, Chugai Pharmaceutical Co., Ltd, a Japanese corporation ("**Chugai**") or its subsidiaries (if any) shall not be deemed as Affiliates of GNE unless GNE provides written notice to Kiniksa of its desire to include Chugai or its respective subsidiaries (as applicable) as Affiliate(s) of GNE.

1.2 *Agreement*

The term "**Agreement**" shall mean this document including any and all appendices and amendments to it as may be added or amended from time to time in accordance with the provisions of this Agreement.

1.3 *Agreement Term*
The term "**Agreement Term**" shall mean the period of time commencing on the Effective Date and, unless this Agreement is terminated sooner as provided in Article 18 (Term and Termination), expiring on the date when no royalty or other payment obligations under this Agreement are or will become due.

1.4 *Alliance Manager*
The term "**Alliance Manager**" has the meaning set forth in Section 5.9 (Alliance Manager).

1.5 *Antibody*
The term "**Antibody**" shall mean any immunoglobulin molecule [***] whether in [***] or any other [***] form, and will include (a) any [***] (b) any [***], and (c) any [***].

1.6 *Antitrust Clearance Date*
The term "**Antitrust Clearance Date**" shall mean the earliest date on which all applicable waiting periods and approvals required under Antitrust Laws in the United States with respect to the transactions contemplated under this Agreement have expired or have been terminated (in the case of waiting periods) or been received (in the case of approvals), in each case, without the imposition of any conditions.

1.7 *Antitrust Filing*
The term "**Antitrust Filing**" shall mean filings by Kiniksa and GNE with the United States Federal Trade Commission ("**FTC**") and the United States Department of Justice ("**DOJ**") and any other applicable Governmental Authority in the Territory, as required under any Antitrust Laws with respect to the transactions contemplated under this Agreement, together with all required documentary attachments thereto.

1.8 *Antitrust Laws*
The term "**Antitrust Laws**" shall mean the HSR Act and any and all other Applicable Laws designed to prohibit, restrict, or regulate actions for the purpose or effect of monopolization or restraint of trade.

1.9 *APA Breach Claim*
The term "**APA Breach Claim**" has the meaning set forth in Section 14.2.3 (GNE Opportunity to Cure APA Breach Claims on Kiniksa's Behalf).

1.10 *APA Milestone Shortfall*
The term "**APA Milestone Shortfall**" has the meaning set forth in Section 9.3.1 (APA Milestone Shortfall).

1.11 *[***] Forecast*
The term "[***] **Forecast**" has the meaning set forth in Section 4.5.2 (Content of Reports).

1.12 *Applicable Law*
The term "**Applicable Law**" shall mean any law, statute, ordinance, code, rule or regulation that has been enacted by a Governmental Authority (including any Regulatory Authority) and is in force as of the Execution Date or comes into force as of the Effective Date or during the Agreement Term, in each case, to the extent that the same is applicable to the performance by the Parties of their respective obligations under this Agreement.

1.13 *Bankruptcy Code*
The term "**Bankruptcy Code**" has the meaning set forth in Section 19 (Bankruptcy).

1.14 *Bioequivalent or Bioequivalence*
The term "**Bioequivalent**" or "**Bioequivalence**" has the meaning set forth in Section 9.5.5 (Competing Drugs).

1.15 *Biogen*
The term "**Biogen**" has the meaning set forth in Section 1.16 (Biogen APA).

1.16 *Biogen APA*
The term "**Biogen APA**" shall mean that certain Asset Purchase Agreement between Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company ("**Kiniksa Parent**"), and Biogen MA Inc., a Massachusetts corporation ("**Biogen**"), dated as of September 7, 2016, as amended by Amendment No.1 to Asset Purchase Agreement, dated as of July 31, 2017, and as further amended by Amendment No.2 to Asset Purchase Agreement, effective as of the Effective Date (the "**Biogen APA Amendment**").

1.17 *Biogen APA Amendment*
The term "**Biogen APA Amendment**" has the meaning set forth in Section 1.16 (Biogen APA).

1.18 *Biogen Background IP Rights*
The term "**Biogen Background IP Rights**" means (a) the Licensed Patent Rights licensed to Kiniksa under the Biogen APA as Background Licensed Patent Rights (as defined in the Biogen APA); and (b) the intellectual property rights sublicensed to Kiniksa under the Biogen APA as Background Sublicensed Intellectual Property (as defined in the Biogen APA).

1.19 *Biogen Reversion*
The term **"Biogen Reversion"** shall mean the occurrence of a termination of the Biogen APA pursuant to Section 8.2.1, Section 8.2.2, or Section 8.2.3 of the Biogen APA.

1.20 *Biogen Side Letter*
The term **"Biogen Side Letter"** shall mean that certain letter agreement entered into as of the Execution Date by and among GNE, Kiniksa and Biogen.

1.21 *BLA*
The term "**BLA**" shall mean a Biologics License Application, or similar application for marketing approval of the Licensed Products for use in the Field submitted to the FDA, or a foreign equivalent of the FDA.

1.22 *Breaching Party*
The term "**Breaching Party**" has the meaning set forth in Section 18.2.1 (Termination for Breach).

1.23 *Business Day*
The term "**Business Day**" shall mean 9:00 am to 5:00 pm local time on a day other than a Saturday, Sunday or bank or other public or federal holiday in New York, California, or Basel, Switzerland.

1.24 *Calendar Quarter*
The term "**Calendar Quarter**" shall mean each period of three (3) consecutive calendar months, ending March 31, June 30, September 30, and December 31; *provided* that the first Calendar Quarter will begin on the Effective Date and the final Calendar Quarter will end on the last day of the Agreement Term.

1.25 *Calendar Year*
The term "**Calendar Year**" shall mean the period of time beginning on January 1 and ending December 31; *provided* that the first Calendar Year will begin on the Effective Date and end on December 31, and the final Calendar Year will end on the last day of the Agreement Term.

1.26 *Chairperson*
The term "**Chairperson**" has the meaning set forth in Section 5.2 (Members).

1.27 *Change of Control*
The term "**Change of Control**" shall mean, with respect to a Party, that: (a) any Third Party acquires directly or indirectly the beneficial ownership of any voting security of such Party, or if the percentage ownership of such Third Party in the voting securities of such Party is increased through stock redemption, cancellation, or other recapitalization, and immediately after such acquisition or increase such Third Party is, directly or indirectly, the beneficial owner of voting securities representing more than fifty percent (50%) of the total voting power of all of the then outstanding voting securities of such Party; (b) any merger, consolidation, recapitalization, or reorganization of such Party is consummated that would result in shareholders or equity holders of such Party immediately prior to such transaction owning fifty percent (50%) or less of the outstanding voting securities of the surviving entity (or its parent entity) immediately following such transaction; (c) the shareholders or equity holders of such Party approve any plan of complete liquidation of such Party, or an agreement for the sale or disposition by such Party of all or substantially all of such Party's assets, in each case, through one or more related transactions, other than to an Affiliate or pursuant to one or more related transactions that would result in shareholders or equity holders of such Party immediately prior to such transaction owning more than fifty percent (50%) of the outstanding voting securities of the surviving entity (or its parent entity) immediately following such transaction; or (d) the sale or transfer to any Third Party, in one or more related transactions, of all or substantially all of such Party's consolidated assets taken as a whole.

1.28 *Chugai*
The term "**Chugai**" has the meaning set forth in Section 1.1 (Affiliate).

1.29 *Clinical Study*
The term "**Clinical Study**" shall mean a Phase II Study or Phase III Study, as applicable.

1.30 *Clinical Supply Fee*
The term "**Clinical Supply Fee**" has the meaning set forth in Section 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Date).

1.31 *Combination Product*
The term "**Combination Product**" shall mean a Licensed Product that includes:

(a) a single pharmaceutical formulation containing as its active ingredients both a Licensed Antibody and one or more other therapeutically or prophylactically active ingredients;

(b) a combination therapy comprised of a Licensed Antibody and one or more other therapeutically or prophylactically active products, priced and sold in a single package containing such multiple products or packaged separately but sold together for a single price; or

(c) a combination therapy comprised of a Licensed Antibody and a Companion Diagnostic, priced and sold in a single package containing such multiple products or packaged separately but sold together for a single price;

in each case, including all dosage forms, formulations, presentations, line extensions, and package configurations. Each of the therapeutically or prophylactically active ingredients or products other than the Licensed Antibody referred to in the foregoing clause (a) or (b) is an "**Other Component**".

1.32 *Commercially Reasonable Efforts*
The term "**Commercially Reasonable Efforts**" shall mean such level of efforts consistent with the efforts GNE or Kiniksa, as applicable, devotes at the same stage of development or commercialization, as applicable, for its own internally developed (whether internally discovered or in-licensed) pharmaceutical products in a similar area with similar market potential, at a similar stage of their product life taking into account the existence of other competitive products in the market place or under development, the proprietary position of the product, the regulatory structure involved, the anticipated profitability of the product and other relevant factors. It is understood that such product potential may change from time to time based upon changing scientific, business and marketing, and return on investment considerations.

1.33 *Companion Diagnostic*
The term "**Companion Diagnostic**" shall mean any product that is used for predicting or monitoring the response of a human being to treatment with a Licensed Product (*e.g.*, device, compound, kit, biomarker or service that contains a component that is used to detect or quantify the presence or amount of an analyte in a body or tissue that affects the pathogens of the disease).

1.34 *Competing Drug*
The term "**Competing Drug**" has the meaning set forth in Section 9.5.5 (Competing Drugs).

1.35 *Compulsory Sublicense*
The term "**Compulsory Sublicense**" shall mean, for a given country or region in the Territory, a license or sublicense of Licensed Patent Rights granted to a Third Party (a **"Compulsory Sublicensee"**) through the order, decree or grant of a Governmental Authority having competent jurisdiction in such country or region, authorizing such Third Party to manufacture, use, sell, offer for sale, import or export a Licensed Product in such country or region.

1.36 *Compulsory Sublicense Compensation*
The term **"Compulsory Sublicense Compensation"** shall mean for a given country or region in the Territory, the compensation paid to GNE and its Affiliates by a Compulsory Sublicensee for its Compulsory Sublicense.

1.37 *Compulsory Sublicensee*
The term "**Compulsory Sublicensee**" has the meaning set forth in Section 1.35 (Compulsory Sublicense).

1.38 *Confidential Information*
The term "**Confidential Information**" shall mean the terms of this Agreement and any and all information (including business or financial information), data or know-how (including Know-How), whether technical or non-technical, oral or written, that is disclosed by one Party or its Affiliates ("**Disclosing Party**") to the other Party or its Affiliates ("**Receiving Party**"). Confidential Information shall not include any information, data or know-how that:

(i) was generally available to the public at the time of disclosure, or becomes available to the public after disclosure by the Disclosing Party other than through fault (whether by action or inaction) of the Receiving Party or its Affiliates,

(ii) can be evidenced by written records to have been already known to the Receiving Party or its Affiliates prior to its receipt from the Disclosing Party,

(iii) is obtained at any time lawfully from a Third Party under circumstances permitting its use or disclosure,

(iv) is developed independently by the Receiving Party or its Affiliates as evidenced by written records other than through knowledge of Confidential Information of the Disclosing Party, or

(v) is approved in writing by the Disclosing Party for release by the Receiving Party.

1.39 *Continuation Election Notice*
The term "**Continuation Election Notice**" shall mean the notice Kiniksa provides to GNE under Section 18.6.1 (Effects of Termination) describing (a) Kiniksa's *bona fide* intentions to continue ongoing development and commercialization of Licensed Product(s); and (b) Kiniksa's request for GNE's continuation of activities during the termination period or transfer of the data, material and information relating to the Licensed Product(s) in accordance with Section 18.6.1 (Effects of Termination).

1.40 *Control*
The term "**Control**" shall mean (as an adjective or as a verb including conjugations and variations such as "**Controls**," "**Controlled**" or "**Controlling**") (a) with respect to Patent Rights or Know-How, the possession by a Party or its Affiliates of the right to transfer or grant a license, sublicense, or other rights to such Patent Rights or Know-How without violating the terms of any agreement or arrangement between such Party and any other party and without violating any Applicable Law; and (b) with respect to proprietary materials, the possession by a Party or its Affiliates of the ability to supply such proprietary materials to the other Party as provided herein without violating the terms of any agreement or arrangement between such Party and any other party and without violating any Applicable Law. Notwithstanding the foregoing, no Patent Right or Know-How will be "Controlled" by either Party or its Affiliates hereunder if such Patent Right or Know-How is owned or in-licensed by a Third Party that becomes an Affiliate of such Party after the Effective Date as a result of a Change of Control of such Party; *provided* that (i) prior to the date of such transaction, neither such Party nor any of its Affiliates had any rights to any such Patent Rights or Know-How, and (ii) any such Patent Rights or Know-How are not used

after the consummation of such Change of Control in the exercise of such Party's rights or performance of its obligations under this Agreement.

1.41 *Cover*
The term "**Cover**" shall mean (as an adjective or as a verb including conjugations and variations such as "**Covered**," "**Coverage**" or "**Covering**") that the developing, making, using, offering for sale, promoting, selling, exporting or importing of a given compound, formulation or product would infringe a patent in the absence of a license under or ownership in such patent or, with respect to a patent application, would infringe a claim included in such patent application if such patent application were to issue as a patent. The determination of whether the developing, making, using, offering for sale, promoting, selling, exporting or importing of a given compound, formulation, process or product is Covered by a particular Patent Right shall be made on a country-by-country basis.

1.42 *Data Subjects*
The term "**Data Subjects**" has the meaning set forth in Section 7.3 (Data Privacy).

1.43 *Decision Period*
The term "**Decision Period**" has the meaning set forth in Section 13.6.2 (GNE Right).

1.44 *Delivery Material*
The term "**Delivery Material**" has the meaning set forth in Section 7.2 (Shipment).

1.45 *Disclosing Party*
The term "**Disclosing Party**" has the meaning set forth in Section 1.38 (Confidential Information).

1.46 *Disposition Transaction*
The term "**Disposition Transaction**" has the meaning set forth in Section 9.5.8 (Disposition of Rights to Payment).

1.47 *DOJ*
The term "**DOJ**" has the meaning set forth in Section 1.7 (Antitrust Filing).

1.48 *DP Resupply*
The term "**DP Resupply**" has the meaning set forth in Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development).

1.49 *Effective Date*
Subject to Section 20.18.3 (Effective Date; Effective Date Representations and Warranties), the term "**Effective Date**" shall mean the later of (a) the date of the last signature of this Agreement, or (b) if an Antitrust Filing is made, the first Business Day immediately following the Antitrust Clearance Date.

1.50 *EMA*
The term "**EMA**" shall mean the European Medicines Agency or any successor agency or authority thereto.

1.51 *Execution Date*
The term "**Execution Date**" shall, subject to the last paragraph of Section 20.18.4 (Provisions Effective on the Execution Date), mean August 2, 2022.

1.52 *Existing Lead Antibody Patent Rights*
The term "**Existing Lead Antibody Patent Rights**" shall mean Existing Licensed Patent Rights that, as of the Effective Date, contain one or more claims that Cover the Lead Antibody.

1.53 *Existing Licensed Patent Rights*
The term "**Existing Licensed Patent Rights**" shall mean the Patent Rights listed on Appendix 14.1.4(a).

1.54 *FDA*
The term "**FDA**" shall mean the Food and Drug Administration of the United States of America or any successor agency or authority thereto.

1.55 *FDCA*
The term "**FDCA**" shall mean the Food, Drug and Cosmetics Act, as amended.

1.56 *Field*
The term "**Field**" shall mean all uses.

1.57 *First Commercial Sale*
The term "**First Commercial Sale**" shall mean, with respect to any Licensed Product in any country in the Territory, the first sale, transfer or disposition for value to an end user of that Licensed Product in that country after Marketing Authorization for that Licensed Product has been received in that country; *provided*, that, the following shall not constitute a First Commercial Sale: (a) any sale to an Affiliate or Sublicensee (unless the Affiliate or Sublicensee is the last entity in the distribution chain of the Licensed Product), or (b) any transfers of a Licensed Product without consideration or for nominal consideration for use in any Clinical Study, or for any *bona fide* charitable, compassionate use or indigent patient program purpose where Licensed Products are sold at or below cost of goods sold or as a sample.

1.58 *Force Majeure Event*
The term "**Force Majeure Event**" shall mean an event beyond the reasonable control of the affected Party and not caused by the fault or negligence of such Party that prevents or substantially interferes with or delays the performance by such Party of any of its obligations hereunder, which may include an embargo, war, act of war (whether war be declared or not), act of terrorism, insurrection, riot, civil commotion, strike, lockout or other labor disturbance, fire, flood, earthquake, epidemic, pandemic (including the Covid-19 pandemic) or other act of God or act, omission or delay in acting by any Governmental Authority.

1.59 *FTC*
The term "**FTC**" has the meaning set forth in Section 1.7 (Antitrust Filing).

1.60 *FTC Letter*
The term "**FTC Letter**" has the meaning set forth in Section 1.6 (Antitrust Clearance Date).

1.61 *Fundamental Representations*
The term "**Fundamental Representations**" shall mean the representations and warranties of Kiniksa under Section 14.1.1 (Safety Data), Section 14.1.4 (Licensed IP), Section 14.1.5 (Inventors), Section 14.1.6 (Grants), Section 14.1.7 (Authorization), Section 14.1.8 (Control of Licensed IP), Section 14.1.9 (Control of In Progress PN Study Data), Section 14.1.10 (Validity of Patent Rights), Section 14.1.11 (Ownership and Legitimacy of Know-How, Kiniksa Cell Line Materials and Inventory), Section 14.1.12 (Existing Clinical Supply of Lead Product), Section 14.1.14 (DP Resupply), and Section 14.2.1 (Biogen APA).

1.62 *Genentech*
The term "**Genentech**" has the meaning set forth on the cover page.

1.63 *GNE*
The term "**GNE**" has the meaning set forth on the cover page.

1.64 *GNE Caused Required Date Failure*
The term **"GNE Caused Required Date Failure"** has the meaning set forth in Section 5.6.3(b) (Failure to Reach Consensus; Limited Escalation).

1.65 *GNE Group*
The term "**GNE Group**" shall mean collectively GNE, its Affiliates and its Sublicensees.

1.66 *GNE Indemnitees*
The term "**GNE Indemnitees**" has the meaning set forth in Section 15.2 (Indemnification by Kiniksa).

1.67 *GNE Invention*
The term "**GNE Invention**" has the meaning set forth in Section 1.87 (Invention).

1.68 *GNE Know-How*
The term "**GNE Know-How**" shall mean all Know-How that GNE or its Affiliates Control on the effective date of termination of this Agreement that is necessary for, and has prior to the date of such termination actually been utilized by or on behalf of GNE or its Affiliates, for the discovery, manufacture, development or commercialization of the Reversion Product(s).

1.69 *GNE Patent Rights*
The term "**GNE Patent Rights**" shall mean all Patent Rights Covering the GNE Know-How.

1.70 *Governmental Authority*
The term "**Governmental Authority**" shall mean any court, agency, department, authority, tribunal, or other instrumentality of any supra-national, national, state, provincial, county, city, or other political subdivision. For clarity, Governmental Authorities include all Regulatory Authorities.

1.71 *Handle*
The term "**Handle**" shall mean preparing, filing, prosecuting (including interferences, reissue, re-examination, post-grant reviews, *inter-partes* reviews, derivation proceedings and opposition proceedings) and maintaining.

1.72 *HSR Act*
The term "**HSR Act**" shall mean the Hart-Scott-Rodino Antitrust Improvements Act.

1.73 *IFRS*
The term "**IFRS**" shall mean International Financial Reporting Standards.

1.74 *In Progress PN Study*
The term "**In Progress PN Study**" shall mean the Clinical Study for the Lead Antibody covered by and as described in the ClinicalsTrials.gov identifier NCT03816891.

1.75 *In Progress PN Study Data*
The term "**In Progress PN Study Data**" shall mean all data and information Controlled by Kiniksa or its Affiliates that is generated by, or on behalf of, Kiniksa, in the conduct of the In Progress PN Study.

1.76 *IND*
The term "**IND**" shall mean an investigational new drug application as defined in the FDCA and applicable regulations promulgated by the FDA, or the equivalent application to the equivalent agency in any other country or group of countries (*e.g.*, outside the US, Clinical Trial Applications (CTAs)), the filing of which is necessary to commence clinical testing of the Licensed Products in humans.

1.77 *IND Transfer Request Date*
The term "**IND Transfer Request Date**" shall mean the date that GNE in writing requests Kiniksa to transfer regulatory responsibility for the In Progress PN Study to GNE, which date shall be no later than [***] days following Kiniksa's completion of the In Progress PN Study (including completion of the associated final clinical study report therefor).

1.78 *Indemnified Party*
The term "**Indemnified Party**" has the meaning set forth in Section 15.3 (Procedure).

1.79 *Indemnifying Party*
The term "**Indemnifying Party**" has the meaning set forth in Section 15.3 (Procedure).

1.80 *Indication*
The term "**Indication**" shall mean any human indication, disease or condition in the Field that can be treated, prevented, cured or the progression of which can be delayed, excluding an expansion of label claim for an already approved indication. Examples of label expansion include [***]. However, the following shall apply: (a) [***] will be considered as separate Indications; and (b) [***] will be considered a subpopulation of [***] such that in the event that a milestone event is achieved for either [***] or [***], both [***] and [***] will be considered a single Indication.

1.81 *Infringement*
The term "**Infringement**" has the meaning set forth in Section 13.6.1 (Notice).

1.82 *Infringement Response*
The term "**Infringement Response**" has the meaning set forth in Section 13.6.2 (GNE Right).

1.83 *Initial Drug Supply*
The term "**Initial Drug Supply**" has the meaning set forth in Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development).

1.84 *Initiating Party*
The term "**Initiating Party**" has the meaning set forth in Section 13.6.4 (Enforcement Costs and Allocation of Recovery).

1.85 *Initiation*
The term "**Initiation**" shall mean, with respect to a Clinical Study, the first date that a subject (healthy volunteer or patient) is first dosed with the applicable Licensed Product in such Clinical Study.

1.86 *Insolvency Event*
The term "**Insolvency Event**" shall mean circumstances under which a Party (i) has a receiver or similar officer appointed over all or a material part of its assets or undertaking; (ii) passes a resolution for winding-up (other than a winding-up for the purpose of, or in connection with, any solvent amalgamation or reconstruction) or a court makes an order to that effect or a court makes an order for administration (or any equivalent order in any jurisdiction); (iii) enters into any composition or arrangement with its creditors (other than relating to a solvent restructuring); or (iv) ceases to carry on business.

1.87 *Invention*
The term "**Invention**" shall mean an invention that is conceived or reduced to practice in connection with any activity carried out pursuant to this Agreement. Under this definition, an Invention may be conceived by employees of Kiniksa solely or jointly with a Third Party (a "**Kiniksa Invention**"), by employees of the GNE Group solely or jointly with a Third Party (a "**GNE Invention**"), or jointly by employees of Kiniksa and employees of the GNE Group with or without a Third Party (a "**Joint Invention**"). For clarity, inventorship will be determined in accordance with the last sentence of the first paragraph of Section 13.1 (Ownership of Inventions), and to the extent applicable thereunder, Joint Inventions may include any Inventions conceived jointly by employees of Kiniksa and employees of the GNE Group, whether the conception by employees of Kiniksa occurred prior to the Effective Date or during the Agreement Term.

1.88 *Inventory*
The term "**Inventory**" shall mean all existing clinical and non-clinical grade drug product and drug substance, active pharmaceutical ingredient, intermediates and raw materials associated with Licensed Antibodies in the Control of Kiniksa (or its Affiliates), as well as any other existing Materials (such as reference standards and retention samples, critical assay reagents and cell banks), and packaging Controlled by Kiniksa (or its Affiliates) and associated with such Licensed Antibodies and Licensed Products containing therein; *provided*, *however*, Inventory does not include the drug substance, drug product and placebos to be supplied by Kiniksa in accordance with Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody), including for the In Progress PN Study, the Initial Drug Supply and the DP Resupply, and any other supply pursuant to Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications).

1.89 *Joint Invention*
The term "**Joint Invention**" has the meaning set forth in Section 1.87 (Invention).

1.90 *Joint Know-How*
The term "**Joint Know-How**" shall mean Know-How that is made jointly by employees of Kiniksa or its Affiliates and the GNE Group, with or without a Third Party, in connection with any activity carried out pursuant to this Agreement, including all Joint Inventions.

1.91 *Joint Patent Rights*
The term "**Joint Patent Rights**" shall mean all Patent Rights Covering a Joint Invention.

1.92 *JTC*
The term "**JTC**" shall mean the joint transition committee described in Article 5 (Governance).

1.93 *Kiniksa*
The term "**Kiniksa**" shall have the meaning set forth on the cover page.

1.94 *Kiniksa Cell Line Materials*
The term **"Kiniksa Cell Line Materials"** shall mean the cell lines and cell banks currently used or held for use by or on behalf of Kiniksa (or its Affiliates) to manufacture or produce the Lead Antibody.

1.95 *Kiniksa Development Activities*
The term "**Kiniksa Development Activities**" shall have the meaning set forth in Section 4.2 (Development by Kiniksa).

1.96 *Kiniksa Development Plan*
The term "**Kiniksa Development Plan**" shall mean the written development plan for the conduct of the Kiniksa Development Activities attached as Appendix 1.96, as it may be amended in accordance with this Agreement.

1.97 *Kiniksa Indemnitees*
The term "**Kiniksa Indemnitees**" shall have the meaning set forth in Section 15.1 (Indemnification by GNE).

1.98 *Kiniksa Invention*
The term "**Kiniksa Invention**" shall have the meaning set forth in Section 1.87 (Invention).

1.99 *Kiniksa Parent*
The term "**Kiniksa Parent**" shall have the meaning set forth in Section 1.16 (Biogen APA).

1.100 *Kiniksa Transition Activities*
The term "**Kiniksa Transition Activities**" shall mean Kiniksa's obligations to (a) complete the Kiniksa Development Activities in accordance with Section 4.2 (Development by Kiniksa), (b) conduct the technology and Inventory and Kiniksa Cell Line Materials transfer activities in accordance with Article 7 (Technology and Material Transfer), and (c) manufacture or have manufactured, or otherwise supply the clinical supply of Lead Antibody in accordance with Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody).

1.101 *Know-How*
The term "**Know-How**" shall mean data, knowledge and information, including materials, samples, chemical manufacturing and control data, toxicological data, pharmacological data,

preclinical and clinical data, assays, platforms, formulations, specifications, processes, and quality control testing data, that are confidential.

1.102 *Knowledge*
The term "**Knowledge**" shall mean, with respect to Kiniksa, [***].

1.103 *Lead Antibody*
The term "**Lead Antibody**" shall have the meaning set forth in the Recitals.

1.104 *Licensed Antibody*
The term "**Licensed Antibody**" shall mean (a) the Lead Antibody or BIIB22G11; (b) any other Antibody that is, as of the Effective Date, Covered by one or more claims in the Existing Lead Antibody Patent Rights; or (c) [***].

1.105 *Licensed IP*
The term "**Licensed IP**" shall mean the Licensed Know-How and Licensed Patent Rights.

1.106 *Licensed Know-How*
The term "**Licensed Know-How**" shall mean all Know-How Controlled by Kiniksa, except the Joint Know-How, that is (a) as of the Effective Date, necessary or reasonably useful, or (b) following the Effective Date during the Agreement Term, necessary, in each case (both (a) and (b)) to develop, have developed, make, have made, use, have used, register, have registered, perform medical affairs with respect to, sell, have sold, offer for sale, import, or export Licensed Antibodies or Licensed Products in the Field.

1.107 *Licensed Patent Rights*
The term "**Licensed Patent Rights**" shall mean all Patent Rights Controlled by Kiniksa, other than the Joint Patent Rights, as of the Effective Date or during the Agreement Term that claim or disclose the Licensed Know-How, and includes the Existing Licensed Patent Rights.

1.108 *Licensed Product*
The term "**Licensed Product**" shall mean any product, including any Combination Product, that contains or incorporates a Licensed Antibody. [***]

1.109 *Marketing Authorization*
The term **"Marketing Authorization"** shall mean, on a country-by-country basis, for a given Licensed Product, the regulatory approval required by Applicable Law to sell such Licensed Product in a country or region in the Territory. For example, (a) "Marketing Authorization" in the United States means final approval of an NDA, sNDA, BLA or sBLA permitting marketing of such Licensed Product in interstate commerce in the United States; (b) "Marketing Authorization" in Europe means marketing authorization for such Licensed Product granted either by a Regulatory Authority in any country in Europe or by the EMA pursuant to Council Directive 2001/83/EC, as amended, or Council Regulation 2309/93/EEC, as amended.

1.110 *Materials*
The term "**Materials**" shall mean any materials relating to the Licensed Antibody or Licensed Product, including chemical or biological substances such as: (i) vector or construct, whether plasmid, phage, virus or any other type; (ii) host organism, including bacteria and eukaryotic cells; (iii) eukaryotic or prokaryotic cell line or expression system; (iv) protein, including any peptide or amino acid sequence, enzyme, antibody or protein conferring targeting properties

and any fragment of a protein or peptide or enzyme; (v) genetic material, including any genetic control element (e.g., promoters); (vi) assay or reagent or other raw materials; or (vii) human biological samples from Clinical Studies conducted by Kiniksa for Licensed Antibody or Licensed Product.

1.111 *Member*
The term "**Member**" shall have the meaning set forth in Section 5.2 (Members).

1.112 *Negotiation Period*
The term "**Negotiation Period**" has the meaning set forth in Section 9.5.8 (Disposition of Rights to Payment).

1.113 *Net Sales*
The term "**Net Sales**" shall mean, for a Licensed Product in a particular period, (a) the amount calculated by subtracting from the Sales of such Licensed Product for such period: (i) a lump sum deduction of [***] of Sales in lieu of those deductions that are not accounted for on a Licensed Product-by-Licensed Product basis (*e.g.*, freight, postage charges, transportation insurance, packing materials for dispatch of goods, custom duties); (ii) uncollectible amounts accrued during such period based on a proportional allocation of the total bad debts accrued during such period and not already taken as a gross-to-net deduction in accordance with the then currently used IFRS in the calculation of Sales of such Licensed Product for such period; (iii) credit card processing fees accrued during such period on such Sales and not already taken as a gross-to-net deduction in accordance with the then currently used IFRS in the calculation of Sales of such Licensed Product for such period; and (iv) government mandated fees and taxes and other government charges accrued during such period not already taken as a gross-to-net deduction in accordance with the then currently used IFRS in the calculation of Sales of such Licensed Product for such period, including, for example, any fees, taxes or other charges that become due in connection with any healthcare reform, change in government pricing or discounting schemes, or other action of a government or regulatory body; and (b) any Compulsory Sublicense Compensation received by the GNE Group in such period. To the extent any of the deduction classes specified in clause (a) above are costs applicable to more than one GNE product, GNE shall fairly allocate such costs among such products in a manner consistent with its internal cost accounting practices. For clarity, no deductions taken in calculating Sales under Section 1.142 (Sales) may be taken a second time in calculating Net Sales.

1.114 *Non-Breaching Party*
The term "**Non-Breaching Party**" shall have the meaning set forth in Section 18.2.1 (Termination for Breach).

1.115 *OSMR*
The term "**OSMR**" shall mean the oncostatin M receptor, one of the receptor proteins for oncostatin M that in humans is encoded by the OSMR gene.

1.116 *Party*
The term "**Party**" shall mean Kiniksa or GNE, as the case may be, and "**Parties**" shall mean Kiniksa and GNE collectively.

1.117 *Patent Rights*
The term "**Patent Rights**" shall mean the rights and interests in and to issued patents and pending patent applications (which, for purposes of this Agreement, include certificates of invention, applications for certificates of invention and priority rights) in any country or region, including any divisionals, continuations, continuations-in-part, substitutions, patents of addition, reissues, extensions, re-examinations or renewal applications related to, or claiming priority to, the foregoing (including any supplemental patent certificates) or any confirmation patent or registration patent, and all patents issuing on, and all foreign counterparts of, any of the foregoing.

1.118 *Payment Currency*
The term "**Payment Currency**" has the meaning set forth in Section 10.3 (Method of Payment).

1.119 *Payment Rights*
The term "**Payment Rights**" has the meaning set forth in Section 9.5.8 (Disposition of Rights to Payment).

1.120 *Peremptory Notice Period*
The term "**Peremptory Notice Period**" has the meaning set forth in Section 18.2.1 (Termination for Breach).

1.121 *Pharmacovigilance Agreement*
The term "**Pharmacovigilance Agreement**" shall mean an agreement entered into by the Parties to set forth the responsibilities and obligations of the Parties with respect to the procedures and timeframes for compliance with Applicable Laws pertaining to safety of the Licensed Product and its related activities.

1.122 *Phase II Study*
The term "**Phase II Study**" shall mean a human clinical trial conducted in any country that would satisfy the requirements of 21 CFR 312.21(b) and is intended to explore one or more doses, dose response, and duration of effect, and to generate initial evidence of clinical activity and safety, for any product in the target patient population. For clarity, a Phase IIa or Phase IIb clinical trial shall be classified as a "Phase II Study."

1.123 *Phase III Study*
The term "**Phase III Study**" shall mean a human clinical trial in an extended human patient population designed to obtain data determining efficacy and safety of any product to support Marketing Authorizations in the proposed therapeutic Indication, as more fully defined in 21 C.F.R. §312.21(c), or its successor regulation, or the equivalent in any foreign country. For clarity, a Phase IIIa or Phase IIIb clinical trial shall be classified as a "Phase III Study."

1.124 *PII/Samples*
The term "**PII/Samples**" has the meaning set forth in Section 18.6.3.3 (Limitations on Grant-Backs; Transfer Expenses).

1.125 *PN Study Know-How*
The term "PN Study Know-How" has the meaning set forth in Section 14.1.8 (Control of Licensed IP).

1.126 *Publishing Notice*
The term "**Publishing Notice**" has the meaning set forth in Section 17.5 (Publications).

1.127 *Quality Agreement*
The term "**Quality Agreement**" has the meaning set forth in Section 6.1.3 (Quality Agreement).

1.128 *Receiving Party*
The term "**Receiving Party**" has the meaning set forth in Section 1.38 (Confidential Information).

1.129 *Regulatory Authority*
The term "**Regulatory Authority**" shall mean any national, supranational (*e.g.*, the European Commission, the Council of the European Union, the European Medicines Agency), regional, state or local regulatory agency, department, bureau, commission, council or other governmental entity with authority over the distribution, importation, exportation, manufacture, production, use, storage, transport, clinical testing, marketing, pricing or sale of a Licensed Product, including the FDA, the EMA, and the European Commission.

1.130 *Regulatory Exclusivity*
The term "**Regulatory Exclusivity**" shall mean, with respect to any Licensed Product in any country or jurisdiction in the Territory, the period of time during which: (a) a Party or its Affiliate or sublicensee has been granted any exclusive marketing rights, other than a Patent Right, by any Governmental Authority under Applicable Law that prevents Third Parties from selling such Licensed Product in such country or jurisdiction, including orphan drug exclusivity, pediatric exclusivity, rights conferred in the US under the FDCA, in the European Union under Directive 2001/83/EC, or rights similar thereto in other countries or regulatory jurisdictions in the Territory; or (b) the data and information submitted by a Party or its Affiliate or sublicensee to the relevant Regulatory Authority in such country or jurisdiction for purposes of obtaining Marketing Authorization of such Licensed Product may not be disclosed, referenced, or relied upon in any way by any Third Party or such Regulatory Authority to support Marketing Authorization of any product by any Third Party in such country or jurisdiction, or if such data and information is disclosed, referenced, or relied upon to support a Marketing Authorization granted to any Third Party in such country or jurisdiction, then such product may not be placed on the market for any indication.

1.131 *Regulatory Filing*
The term **"Regulatory Filing**" shall mean, collectively: (a) any IND, CTA, Marketing Authorization Application, BLA, sBLA, establishment license application, drug master file, application for designation as an "Orphan Drug" under the Orphan Drug Act, for "Fast Track" status under Section 506 of the FDCA (21 U.S.C. § 356) or for a Special Protocol Assessment under Section 505(b)(4)(B) and (C) of the FDCA (21 U.S.C. § 355(b)(4)(B)) and all other similar filings (including counterparts of any of the foregoing in any country or region in the Territory); (b) all supplements and amendments to any of the foregoing; and (c) all data and other information contained in, and correspondence relating to, any of the foregoing.

1.132 *Rep Failure*
The term "**Rep Failure**" has the meaning set forth in Section 20.18.3 (Effective Date; Effective Date Representations and Warranties).

1.133 *Report*
The term "**Report**" has the meaning set forth in Section 4.5.1 (Records; Reports).

1.134 *Residual WCB*
The term **"Residual WCB"** has the meaning set forth in Section 6.1.2.3 (Residual WCB).

1.135 *Reversion Negotiation Expiration*
The term **"Reversion Negotiation Expiration"** has the meaning set forth in Section 18.6.1(d) (Effects of Termination).

1.136 *Required Supply Date*
The term "**Required Supply Date**" has the meaning set forth in Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development).

1.137 *Reversion Product*
The term "**Reversion Product**" has the meaning set forth in Section 18.6.1 (Effects of Termination).

1.138 *Roche*
The term "**Roche**" has the meaning set forth on the cover page.

1.139 *ROFN Exercise Period*
The term "**ROFN Exercise Period**" has the meaning set forth in Section 9.5.8 (Disposition of Rights to Payment).

1.140 *ROFN Notice*
The term "**ROFN Notice**" has the meaning set forth in Section 9.5.8 (Disposition of Rights to Payment).

1.141 *Royalty Term*
The term "**Royalty Term**" shall mean, with respect to a given Licensed Product and for a given country, the period of time commencing on the date of First Commercial Sale of the Licensed Product in such country and ending on the latest of (a) the expiration of the last to expire Valid Claim within the [***] that Covers the [***] of such Licensed Product in such country; (b) the expiration of Regulatory Exclusivity in such country, and (c) ten (10) years from the date of the First Commercial Sale of such Licensed Product in such country.

1.142 *Sales*
The term "**Sales**" shall mean, for a Licensed Product in a particular period, the sum of (i) and (ii):

(i) the amount stated in the Roche Holding AG "Sales" line of its externally published audited consolidated financial statements with respect to such Licensed Product for such period (excluding sales to any Sublicensees that are not Affiliates of GNE, or by any Compulsory Sublicensee). This amount reflects the gross invoice price at which such Licensed Product was sold or otherwise disposed of (other than for use as clinical supplies or free samples) by GNE and its Affiliates to such Third Parties (excluding sales to any Sublicensees that are not Affiliates of GNE or to any Compulsory Sublicensee) in such period reduced by gross-to-net deductions, if not previously deducted from such invoiced amount, taken in accordance with the then currently used IFRS.

By way of example, the gross-to-net deductions taken in accordance with IFRS as of the Effective Date include items such as the following:

[***]

(ii) for Sublicensees that are not GNE Affiliates [***], the sales amounts reported to GNE and its Affiliates in accordance with the sublicense contractual terms and their then-currently used accounting standards. For the purpose of clarity, any such Sublicensee sales as reported to GNE in accordance with [***] shall be excluded from the sales amount.

1.143 *Settlement*
The term "**Settlement**" has the meaning set forth in Section 13.6.6 (Settlement).

1.144 *Sublicensee*
The term "**Sublicensee**" shall mean a person or entity to which GNE has licensed rights (through one or multiple tiers), other than a Compulsory Sublicensee, pursuant to this Agreement.

1.145 *Subsequent Clinical Study-Related Contracts*
The term "**Subsequent Clinical Study-Related Contracts**" has the meaning set forth in Section 7.4.1 (Subsequent Clinical Study-Related Contracts).

1.146 *Suit Notice*
The term "**Suit Notice**" has the meaning set forth in Section 13.6.2 (GNE Right).

1.147 *Tech Transfer*
The term "**Tech Transfer**" shall mean the transfer of the Transfer Know-How and Materials to GNE pursuant to Article 7 (Technology and Material Transfer).

1.148 *Tech Transfer Plan*
The term "**Tech Transfer Plan**" shall mean the plan for transfer of the Transfer Know-How and Materials to GNE set forth on Appendix 1.148, as it may be amended in accordance with this Agreement.

1.149 *Territory*
The term "**Territory**" shall mean all countries of the world.

1.150 *Third Party*
The term "**Third Party**" shall mean a person or entity other than (i) Kiniksa or any of its Affiliates or (ii) GNE or any other member of the GNE Group.

1.151 *Transfer Know-How and Materials*
The term "**Transfer Know-How and Materials**" has the meaning set forth in Section 7.1.1 (Transfer Obligations).

1.152 *US*
The term "**US**" shall mean the United States of America and its territories and possessions.

1.153 *US$*
The term "**US$**" shall mean US dollars.

1.154 *Valid Claim*
The term "**Valid Claim**" shall mean (a) an issued and unexpired claim within the Licensed Patent Rights or the Joint Patent Rights, in each case, that has not been disclaimed, revoked or held invalid by a final non-appealable decision of a court of competent jurisdiction or government agency or (b) a claim of a pending patent application within the Licensed Patent Rights or the Joint Patent Rights that, in the case of any such patent application, was filed in good faith, has not been pending for more than [***] and has not been abandoned or finally disallowed.

2. Grant of License

2.1 *Licenses*
Subject to the terms and conditions of this Agreement, Kiniksa hereby grants to GNE an exclusive (even as to Kiniksa, subject to Section 2.3 (No Implied Rights; Retained Rights)) right and license, including the right to sublicense (through multiple tiers), under Kiniksa's interest in the Joint Know-How, the Joint Patent Rights, and the Licensed IP to research, have researched, develop, have developed, register, have registered, use, have used, make, have made, import, have imported, export, have exported, market, have marketed, distribute, have distributed, sell and have sold Licensed Antibodies and Licensed Products in the Field in the Territory. Notwithstanding any provision to the contrary set forth in this Agreement, nothing in the foregoing is intended to grant to GNE any rights from Kiniksa in any Antibody that is Controlled by Kiniksa other than Licensed Antibodies.

2.2 *Sublicense*

2.2.1 Right to Sublicense to Affiliates

GNE shall have the right to grant sublicenses to its Affiliates (through multiple tiers), under its rights granted under Section 2.1 (Licenses) without prior approval of Kiniksa. If GNE grants such a sublicense, then GNE shall ensure that all of the applicable terms and conditions of this Agreement shall apply to the Affiliate to the same extent as they apply to GNE for all applicable purposes. The grant of any sublicense in compliance with this Section 2.2.1 (Right to Sublicense to Affiliates) shall not relieve GNE of its obligations under this Agreement, and as between Kiniksa and GNE, GNE assumes full responsibility for the performance of all obligations to be performed by, and observance of all terms so imposed on, such Affiliate.

2.2.2 Right to Sublicense to Third Parties
GNE and its Affiliates shall have the right to grant written sublicenses to non-Affiliate entities (through multiple tiers) under its rights granted under Section 2.1 (Licenses) without prior approval of Kiniksa. GNE shall notify Kiniksa reasonably promptly (and no later than [***] days) after the execution of an agreement under this Section 2.2.2 (Right to Sublicense to Third Parties) (including the identity of the applicable Sublicensee and the region in which such rights have been sublicensed). Any sublicense granted by GNE shall be consistent with the applicable terms and conditions of this Agreement. The grant of any sublicense in compliance with this Section 2.2.2 (Right to Sublicense to Third Parties) shall not relieve GNE of its obligations under this Agreement, and GNE shall be responsible for the payment of all amounts due hereunder and for all other obligations of its Sublicensees under this Agreement as if such obligations were those of GNE.

2.2.3 Right to Subcontract

GNE shall have the right to subcontract the work performed under this Agreement without the prior approval of Kiniksa. Any subcontract entered into by GNE shall be consistent with the applicable terms and conditions of this Agreement. GNE shall be responsible for the actions or omissions of its subcontractors in performing work hereunder and the compliance of its subcontractors with the terms and conditions of this Agreement.

2.3 *No Implied Rights; Retained Rights*

Except as specifically set forth in this Agreement, this Agreement shall not be construed as (i) giving any of the Parties any license, right, title, interest in, or ownership to the Confidential Information of the other Party; (ii) granting any license or right under any intellectual property rights; or (iii) representing any commitment by either Party to enter into any additional agreement, by implication or otherwise. Any rights not expressly granted by a Party to the other Party under this Agreement are hereby retained by such Party. Notwithstanding any provision to the contrary set forth in this Agreement, Kiniksa may, and hereby retains the non-exclusive right under the Licensed IP and Kiniksa's interest in the Joint Know-How and the Joint Patent Rights solely to, perform the Kiniksa Transition Activities in the Territory until such time as the Kiniksa Transition Activities have been completed.

3. Diligence

GNE and Kiniksa shall use Commercially Reasonable Efforts to perform their respective activities contemplated by this Agreement. Specifically, GNE agrees to use Commercially Reasonable Efforts to pursue further development and commercialization of Licensed Products in the Field in the Territory.

4. Development and Commercialization

4.1 *Development by GNE*

Subject to Kiniksa's responsibility for conducting the Kiniksa Transition Activities as set forth in Section 4.2 (Development by Kiniksa), Article 7 (Technology and Material Transfer) and Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody), GNE, at its own cost, shall control and be responsible for pursuing all activities initiated after the Effective Date for the development of Licensed Antibodies and Licensed Products in the Field in the Territory.

4.2 *Development by Kiniksa*

Kiniksa shall, at its own cost, use Commercially Reasonable Efforts to continue to conduct and finalize the In Progress PN Study (including completion of the associated final clinical study report therefor), as set forth in and in accordance with the Kiniksa Development Plan (such development activities the "**Kiniksa Development Activities**"). Any proposed modifications to the Kiniksa Development Plan shall be made in accordance with Section 5.3 (Responsibilities of the JTC).

4.3 *Exchange of Information*

On a Calendar Quarter basis, for so long as Kiniksa is performing any Kiniksa Development Activities or other Kiniksa Transition Activities, the Parties shall disclose and make available to each other all data and information necessary for Kiniksa to conduct and report to GNE on the status of the Kiniksa Development Activities and other Kiniksa Transition Activities. The Parties, through the JTC at its regularly scheduled meetings or otherwise as may be reasonably requested by a Member of the JTC, shall answer any questions reasonably posed and provide

any information reasonably requested related to the Kiniksa Development Activities and other Kiniksa Transition Activities.

4.4 *Commercialization*

GNE, at its own cost, shall have sole responsibility and decision-making authority for the marketing, promotion, sale and distribution of Licensed Products in the Territory.

4.5 *Reports to Kiniksa*

4.5.1 Records; Reports.

GNE shall (a) maintain records of its development and commercialization activities with respect to Licensed Antibodies and Licensed Products under this Agreement in sufficient detail and in good scientific manner, which records shall reflect work performed and results achieved in the conduct of such development and commercialization activities and (b) keep Kiniksa reasonably informed through the JTC at its regularly scheduled meetings, regarding the development activities conducted with respect to Licensed Antibodies and Licensed Products during the duration of the JTC in accordance with Section 5.7 (Information Exchange), and thereafter by providing Kiniksa with reports regarding GNE's development and commercialization activities conducted with respect to Licensed Antibodies and Licensed Products at least [***] summarizing the activities undertaken by the GNE Group for the relevant [***] period (each, a "**Report**").

4.5.2 Content of Reports.

Any Reports provided pursuant to Section 4.5.1 (Records; Reports) will include at least information regarding: (a) completed activities with respect to the development of Licensed Antibodies and Licensed Products as well as the anticipated development activities planned in the subsequent [***]; (b) activities with respect to the milestone events described in Section 9.3 (Development and Regulatory Event Payments), including when such milestone events are expected to be achieved and whether or not such milestone events have been achieved; (c) an updated list of the Licensed Patent Rights; and (d) the anticipated date and actual date, as applicable, of the First Commercial Sale of each Licensed Product in each country of the Territory; *provided, however*, that after a Licensed Product receives Marketing Authorization, the information required in (a) will only need to be provided [***] and will include planned activities for the subsequent [***]. In addition, in order to enable Kiniksa to prepare its quarterly and annual public disclosures regarding Kiniksa's results of operations, on a Licensed Product-by-Licensed Product basis, upon the earlier of (i) [***] prior to the anticipated First Commercial Sale of such Licensed Product in any country in the Territory, or (ii) the date of GNE's submission of a BLA for a Licensed Product in any country in the Territory, and on a [***] basis thereafter, GNE shall prepare a commercialization report, which report shall include a non-binding timeline for achieving First Commercial Sale and a non-binding [***] good faith rolling forecast of Net Sales and Sales of Licensed Products in the Field in the Territory, broken down by Calendar Quarters (such forecast, an "[***] **Forecast**"). Thereafter, GNE shall provide to Kiniksa an updated [***] Forecast by [***] of each Calendar Year of the Agreement Term.

4.5.3 Inability to Report Specified Details Not a Material Breach

Notwithstanding any provision to the contrary in Section 4.5.1 (Records; Reports) or Section 4.5.2 (Content of Reports), any failure by GNE to comply with required content or time periods set forth therein will not be considered a material breach of this Agreement if GNE is unable to satisfy the express content for such reports when applying GNE's then-standard processes, practices and operational procedures, so long as GNE provides such information as reasonably and practicably available applying GNE's then-current standards.

5. Governance

5.1 *Joint Transition Committee*
Within [***] Business Days after the Effective Date, the Parties shall establish a joint transition committee ("**JTC**") to oversee the Kiniksa Transition Activities for, and for sharing information relating to the development of, the Licensed Antibodies and the Licensed Products under this Agreement.

5.2 *Members*
The JTC shall be composed of up to [***] persons ("**Members**"). GNE and Kiniksa each shall be entitled to appoint up to [***] Members with appropriate seniority, functional expertise, and decision-making authority. Each Party may replace any of its Members and appoint a person to fill the vacancy arising from each such replacement. A Party that replaces a Member shall notify the other Party at least [***] days prior to the next scheduled meeting of the JTC. Both Parties shall use reasonable efforts to keep an appropriate level of continuity in representation. Both Parties may invite a reasonable number of additional experts or advisors to attend part or the whole JTC meeting with prior notification to the JTC, *provided* that they are subject to confidentiality agreements consistent with the obligations of confidentiality set forth in Article 17 (Confidential Information) of this Agreement. The JTC shall be chaired by a GNE Member ("**Chairperson**").

5.3 *Responsibilities of the JTC*
The JTC shall have the responsibility and authority to:

[***]

The JTC shall have no responsibility or authority other than as expressly set forth in this Section 5.3 (Responsibilities of the JTC).

5.4 *Meetings*
The Chairperson or his/her delegate will be responsible for sending invitations and agendas for all JTC meetings to all Members at least [***] days before the next scheduled meeting of the JTC. The venue for the meetings shall be agreed by the JTC. The JTC shall hold meetings at least [***], either in person or by tele-/video-conference, and in any case as frequently as the Members of the JTC may agree shall be necessary, but not more than [***] unless otherwise agreed by the Parties. The Alliance Manager of each Party may attend the JTC meetings as a permanent participant.

5.5 *Minutes*
The Chairperson will be responsible for designating a Member to record in reasonable detail and circulate draft minutes of JTC meetings to all members of the JTC for comment and review within [***] days after the relevant meeting. The Members of the JTC shall have [***] days to provide comments. The Party preparing the minutes shall incorporate timely received comments and distribute finalized minutes to all Members of the JTC within [***] days of the relevant meeting.

5.6 *Decisions*

5.6.1 Decision Making Authority
The JTC shall have decision-making authority solely over those matters within its purview, as set forth in Section 5.3 (Responsibilities of the JTC).

5.6.2 Consensus; Good Faith
The Members of the JTC shall act in good faith to cooperate with one another and seek agreement with respect to issues to be decided by the JTC. The Parties shall endeavor to make decisions by consensus, with each Party having one vote.

5.6.3 Failure to Reach Consensus; Limited Escalation
If the JTC is unable to decide a matter within its purview by consensus, then GNE shall have the final decision authority on any such matter; *provided* that:

(a) with regard to the obligations, timing of transfer, or process for such transfer of any material or information set forth in the Tech Transfer Plan, GNE shall not have the right to require Kiniksa to change such obligations, timing, or process in a manner that Kiniksa deems in good faith as an unreasonable request (*provided* that Kiniksa agrees to use good faith reasonable effort to accommodate any such change request), and if the JTC fails to reach consensus with respect to such obligations, timing, or process, then no changes will be made and the *status quo* will prevail;

(b) (i) if GNE exercises its final decision-making authority over a matter that Kiniksa has notified GNE may result in a delay to its delivery of the Initial Drug Supply or DP Resupply by the applicable Required Supply Date, then notwithstanding any resulting delay (each a "**GNE Caused Required Date Failure**"), GNE will pay the Clinical Supply Fee to the extent required pursuant to the last sentence of Section 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Dates), *except* that GNE may exercise its final decision-making authority to ensure the Initial Drug Supply and DP Resupply comply with the requirements set forth in the first sentence of Section 6.1.2.5 (Additional Clinical Supply and Specifications) and such exercise of GNE's final decision-making authority will not be deemed a GNE Caused Required Date Failure in such case; and (ii) subject to the foregoing clause (i), if GNE, through its exercise of final decision-making authority, makes any decision relating to drug substance or drug product manufacturing, storage, or testing by Kiniksa for the Lead Antibody (other than changes solely required to ensure that the Initial Drug Supply and DP Resupply comply with the requirements set forth in the first sentence of Section 6.1.2.5 (Additional Clinical Supply and Specifications)) and such decision results in a material increase in Kiniksa's internal or out-of-pocket costs for such activities, then GNE shall reimburse Kiniksa for [***] of such increase in Kiniksa's documented internal and out-of-pocket costs for such activities; or

(c) if such matter relates to an amendment to the then-current Kiniksa Development Plan or otherwise would have a material impact on the In Progress PN Study, then such matter shall be first referred to the Parties' respective Alliance Managers for such Alliance Managers to propose a recommendation for resolution to the JTC within [***] days after such referral, and if the Alliance Managers are unable to agree on a recommendation within such [***] day period or if the JTC does not accept such recommendation within [***] days after the proposal of such recommendation by the Alliance Managers, then either Kiniksa or Genentech may, by written notice to the other Party, refer the matter to the Chief Executive Officer of Kiniksa (or their designee) and a vice president of GNE who has a managerial decision making role over the

disputed issue (or their designee) for resolution, who together shall use reasonable and good faith efforts to reach a decision by consensus within [***] days after the date such matter is referred to them. If the Parties' executives fail to resolve such matter, then, notwithstanding any provision to the contrary set forth in this Agreement, neither Party will have final decision-making authority over such matter and the *status quo* will prevail, except that (i) if such matter would not require Kiniksa to perform additional activities not specified under the Kiniksa Development Plan and (ii) such GNE vice president (or their designee) in good faith concludes that preserving the *status quo* would materially impact GNE's development program for the Licensed Antibodies, then GNE shall have the final decision-making authority on such matter, and if any decision under this clause (c) results in a material increase in Kiniksa's internal or out-of-pocket costs for such activities, then GNE shall reimburse Kiniksa for [***] of such increase in Kiniksa's documented internal and out-of-pocket costs for such activities. Any such decision shall constitute a decision of the JTC.

5.7 *I*nformation Exchange

Kiniksa shall disclose to GNE the information in relation to the Kiniksa Transition Activities through the JTC. The JTC may determine other routes of information exchange.

During the lifetime of the JTC as set forth in Section 5.12 (Lifetime), GNE shall disclose to Kiniksa the information required under Section 4.5 (Reports to Kiniksa) regarding GNE's Licensed Product development activities under this Agreement through regularly scheduled meetings of the JTC. After the lifetime of the JTC, GNE shall provide such information to Kiniksa, as set forth in Section 4.5 (Reports to Kiniksa).

5.8 *Subcommittees*

The JTC shall have the right, within the scope of its responsibilities specified in Section 5.3 (Responsibilities of the JTC), to establish sub-committees or sub-teams; *provided* that any decision making under such sub-committees or sub-teams shall be approved by the JTC.

5.9 *Alliance Manager*

Each Party shall appoint one person to be its point of contact with responsibility for facilitating communication and collaboration between the Parties during the duration of the JTC and the Agreement Term (each, an "**Alliance Manager**"). The Alliance Managers shall be permanent participants of the JTC meetings (but not members of the JTC). The Alliance Managers shall facilitate resolution of potential and pending issues and potential disputes to reach consensus and avert escalation of such issues or potential disputes.

5.10 *Limitations of Decision-Making Authority*

Notwithstanding anything to the contrary set forth in this Agreement, without the other Party's prior written consent, neither GNE (in the exercise of GNE's final decision-making authority), the JTC, any sub-committee, nor a Party's executive officer, in each case, may make a decision that could reasonably be expected to (a) require the other Party to take any action that such other Party reasonably believes would (i) require such other Party to violate any Applicable Law, the requirements of any Regulatory Authority, or any agreement with any Third Party entered into by such other Party prior to the Effective Date, or (ii) require such other Party to infringe or misappropriate any intellectual property rights of any Third Party; or (b) conflict with, amend, interpret, modify, or waive compliance under this Agreement.

5.11 *Expenses*
Each Party shall be responsible for its own expenses, including travel and accommodation costs, incurred in connection with participating in the JTC.

5.12 *Lifetime*
The JTC shall exist commencing from its formation after the Effective Date and shall exist for so long as Kiniksa is performing the Kiniksa Transition Activities. Following the completion of the Kiniksa Transition Activities, the JTC will be permanently disbanded.

6. Manufacture and Supply

6.1 *Clinical Supply of Licensed Product*

6.1.1 GNE Responsibility
Except with respect to Kiniksa's responsibilities for providing certain clinical supply of the Lead Antibody as set forth in Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody), following the completion of the Tech Transfer, GNE shall have the sole right and responsibility, at its own cost, for the manufacture and supply of clinical supplies of Licensed Antibody and Licensed Product.

6.1.2 Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody

6.1.2.1 Kiniksa Clinical Supply for Kiniksa Development Activities
Kiniksa shall be responsible for providing, at its own cost, all clinical supply (including any necessary drug substance, drug product or placebo controls) of the Lead Antibody required for Kiniksa's completion of the In Progress PN Study.

6.1.2.2 Kiniksa Clinical Supply for GNE Development
Kiniksa shall provide to GNE drug product, drug substance, and placebo controls (including in each case related reference standard materials and diluents) for the Lead Antibody consisting of an "**Initial Drug Supply**" and a "**DP Resupply**," as each such term is further defined on, and in accordance with, Appendix 6.1.2.2. The Initial Drug Supply and the DP Resupply shall each conform to the requirements of Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications) and be delivered by Kiniksa to GNE in the quantities specified in Appendix 6.1.2.2 by no later than [***] days after the Effective Date for the Initial Drug Supply and [***] for the DP Resupply (each such date as applicable, the **"Required Supply Date"**); *provided* that Kiniksa shall use Commercially Reasonable Efforts to deliver the DP Resupply by [***]. The Initial Drug Supply and DP Resupply shall be manufactured and supplied by Kiniksa at its own cost but will be subject to a fixed payment subject to and in accordance with Section 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Dates). Any failure by Kiniksa to deliver to GNE the Initial Drug Supply and DP Resupply by the applicable Required Supply Date shall not release Kiniksa from its obligation under this Agreement to supply such Initial Drug Supply and DP Resupply in accordance with the terms of this Agreement; *provided* that, if Kiniksa is unable to deliver the DP Resupply by the applicable Required Supply Date due to one or more batch failures in the manufacture thereof (and, subject to Section 5.6.3(b), such failure results in GNE not being obligated to pay Kiniksa the amount set forth in Section 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Dates)), then at GNE's request, Kiniksa shall complete its obligation to supply such DP Resupply to GNE as soon as practicable and GNE will reimburse Kiniksa's [***] for the manufacture thereof.

6.1.2.3 Residual WCB

Kiniksa shall, or cause its contract manufacturer to, maintain working cell banks reasonably sufficient to fulfill Kiniksa's responsibilities and obligations under Section 6.1.2.1 (Kiniksa Clinical Supply for Kiniksa Development Activities), Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development), and Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications) (the "**Residual WCB**") until such time as Kiniksa has completed its obligations under the foregoing Sections. The amount of Residual WCB is set forth in the Tech Transfer Plan. Thereafter, Kiniksa shall, or cause its contract manufacturer to, deliver any remaining Residual WCB to GNE.

6.1.2.4 Any Remaining Clinical Supply

Promptly following Kiniksa's completion of all of its clinical supply obligations under Section 6.1.2.1 (Kiniksa Clinical Supply for Kiniksa Development Activities), Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development) and if applicable the second paragraph of Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications), subject to Section 6.1.2.3 (Residual WCB), Kiniksa shall deliver, or cause its contract manufacturer to deliver, to GNE all drug substance, drug product, placebos and any Kiniksa Cell Line Materials, including in each case, related reference standard materials, relating to Licensed Antibodies or Licensed Product remaining in Kiniksa's Control.

6.1.2.5 Additional Clinical Supply Obligations and Specifications

The Initial Drug Supply and DP Resupply provided by Kiniksa to GNE under this Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody) shall satisfy the applicable specifications, quality and release requirements for such Initial Drug Supply and DP Resupply as in existence as of the Effective Date (as audited by GNE prior to the Execution Date) and will comply in all material respects with all Good Manufacturing Practices (GMP) and Applicable Laws related thereto. Until delivery by Kiniksa and acceptance by GNE of each of the Initial Drug Supply and the DP Resupply in accordance with Section 6.1.2.2 (Kiniksa Clinical Supply Obligations for GNE Development), Kiniksa shall keep GNE timely informed of any changes to process or manufacturing of drug product and drug substance. Until completion of [***] Kiniksa will [***] and will provide GNE with the data on a rolling basis. After [***], responsibility for [***] shall be transferred to GNE to [***].

Upon GNE's request at any time prior to [***], Kiniksa shall supply an additional batch of drug substance to GNE at Kiniksa's fully burdened manufacturing costs *plus* a markup of [***] percent ([***]%). Kiniksa's "fully burdened manufacturing costs" will include [***]. As between the Parties, GNE will be responsible, at its own cost, for [***]. In addition, Kiniksa shall keep GNE informed, through the JTC, on material decisions relating to drug substance and drug product manufacturing, storage, and testing for Lead Antibody for which Kiniksa has responsibility pursuant to Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody), and the JTC will review, discuss, and determine whether to approve any such decision.

All clinical supply provided by Kiniksa to GNE under Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development), Section 6.1.2.4 (Any Remaining Clinical Supply) or, as applicable, Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications), shall be shipped to GNE in accordance with Section 7.2 (Shipment) and, upon delivery thereof, shall be subject to quality assessment and release procedures conforming with the requirements of this Agreement and the Quality Agreement. In addition, if agreed by the Parties, the Parties shall in good faith

negotiate and enter into a supply agreement to cover the supply obligations of Kiniksa under Section 6.1.2.2 (Kiniksa Clinical Supply for GNE Development) and this Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications).

6.1.3 Quality Agreement

Promptly as practical after the Effective Date, but in no event later than [***] days following the Effective Date, the Parties shall negotiate in good faith and enter into quality agreement(s) (the "**Quality Agreement(s)**") with respect to the supply of drug substance or drug product for the Licensed Product to GNE by, or on behalf or, Kiniksa under this Agreement.

6.2 *Commercial Supply of Licensed Product*

GNE shall be solely responsible, at its own cost, for the commercial manufacture and commercial supply of Licensed Product for sale in the Territory, either by itself or through Third Parties.

7. Technology and Material Transfer

7.1 *Technology and Material Transfer*

7.1.1 Transfer Obligations

Subject to Section 7.1.2 (Newly-Identified Transfer Know-How and Materials) and Section 7.1.3 (Newly-Created Transfer Know-How and Materials), promptly following the Effective Date or at such later time as reasonably determined by GNE, Kiniksa shall initiate a technology transfer to GNE, in accordance with the Tech Transfer Plan, for the transfer of (a) the Licensed Know-How and Inventory existing as of the Effective Date, including Kiniksa Cell Line Materials, documentation, and other Materials within the Licensed IP (such Licensed Know-How, Inventory, Kiniksa Cell Line Materials, documentation and other Materials, the "**Transfer Know-How and Materials"**) and (b) the regulatory data, records, correspondence, and Regulatory Filings relating to the Lead Antibody or Licensed Product, as further detailed in Section 8.2 (Regulatory Transfer), in each case, as set forth in the Tech Transfer Plan. To the extent any copies of documents or materials within the Transfer Know-How and Materials and set forth in the Tech Transfer Plan are in possession of a Kiniksa Affiliate or a Third Party, Kiniksa is solely responsible, at its own cost, to procure and provide to GNE all such Transfer Know-How and Materials, together with all rights to access and use any such Transfer Know-How and Materials in accordance with this Agreement (which may be procured or provided by letters of authorization or comparable instruments where approved by GNE). Kiniksa shall make available its personnel (and use reasonable efforts to cause its Third Party subcontractors with relevant subject matter expertise to be available) on a reasonable basis to consult with GNE with respect to the Transfer Know-How and Materials set forth in the Tech Transfer Plan. Activities of the employees and representatives of each Party in connection with such transfer or any transfer under Section 7.1.2 (Newly-Identified Transfer Know-How and Materials) or Section 7.1.3 (Newly-Created Transfer Know-How and Materials) shall be at such Party's cost.

7.1.2 Newly-Identified Transfer Know-How and Materials

It is understood and agreed that there may be additional Transfer Know-How and Materials that are not identified at the time of the initial transfer of the Transfer Know-How and Materials in accordance with the Tech Transfer Plan but that are thereafter identified. If, after such initial transfer, either Party identifies any such additional Transfer Know-How and Materials that are within the scope of the Licensed IP and in Kiniksa's Control or that would be within the scope of the Licensed IP if Controlled by Kiniksa but that are then Controlled by an Affiliate of Kiniksa,

then Kiniksa shall transfer or cause to be transferred such additional Transfer Know-How and Materials to GNE promptly after such identification and notification.

7.1.3 Newly-Created Transfer Know-How and Materials

It is also understood and agreed that there may be additional Transfer Know-How and Materials that are not yet existing at the time of the initial transfer of Transfer Know-How and Materials as set forth in the Tech Transfer Plan, but are expected to be generated by, or on behalf of, Kiniksa or its Affiliates after the Effective Date and that will constitute Transfer Know-How and Materials. Kiniksa shall provide such additional Transfer Know-How and Materials as soon as reasonably practical after coming into Kiniksa's (or any of its Affiliate's) Control, either in accordance with the timeline as set forth in the Tech Transfer Plan or in reasonable batches, as agreed by the Parties. As an example, Kiniksa expects to generate additional reference standard stability reports, drug substance and drug product stability reports, analytical methods, manufacturing instructions for the process through bulk drug substance and drug product (empty batch records), master batch records ("empty") and executed manufacturing documentations and material safety data sheet(s) for the Inventory, Initial Drug Supply and the DP Resupply transferred to GNE under this Agreement, and, Kiniksa agrees to provide all raw data within [***] after the end of each cohort.

7.2 *Shipment*

Kiniksa shall deliver, or authorize GNE to direct the relevant Kiniksa designee to deliver, to GNE or GNE's designee, all Inventory, the Initial Drug Supply and the DP Resupply, the Residual WCB, and any other material to be shipped to GNE or GNE's designee pursuant to the Tech Transfer Plan, Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody), Section 6.1.2.3 (Residual WCB), Section 6.1.2.4 (Any Remaining Clinical Supply) and if applicable the second paragraph of Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications), as applicable (the "**Delivery Material**"). Unless otherwise specified in this Agreement or as agreed to by the Parties, shipment of Delivery Material from Kiniksa to GNE shall be delivered [***] (Incoterms 2020). Upon receipt of the Delivery Material [***], Kiniksa hereby sells, conveys, assigns and transfers to GNE all of Kiniksa's rights, title, and interests in and to such Delivery Material. Kiniksa shall enter into customary documents, including an appropriate bill of sale, if required, for shipment of Delivery Material to GNE.

7.3 *Data Privacy*

If necessary, the Parties will enter into the relevant agreements under applicable data privacy laws (such as a data transfer agreement) when required. The terms of such agreement will be agreed upon by the Parties when the requirement to enter into such agreement has been confirmed by the Parties. Each Party represents and warrants that (a) any data, including personal data, will be provided to the other Party hereunder in a pseudonymized or de-identified manner as applicable, and that the disclosing Party will not disclose or otherwise make available to the other Party or give access to the other Party to any code allowing identification of data subjects (e.g., patients, study participants, specimen donors) ("**Data Subjects**"); and (b) it has the full right and authority to provide such data to the other Party for the purposes set forth in this Agreement and has taken necessary actions to ensure that all contractual arrangements, notices, and Data Subject consents required by applicable data protection law have been executed, provided, or obtained prior thereto. Furthermore, each Party shall not: (i) undertake any actions to identify the Data Subjects, or (ii) try to get access to any code allowing identification of such Data Subjects.

7.4 *Clinical Transfer*

7.4.1 Subsequent Clinical Study-Related Contracts

As part of the Tech Transfer, Kiniksa shall cooperate with and assist GNE by identifying key Third Party subcontractors engaged by Kiniksa (or its Affiliates) that are performing activities by, for, or with Kiniksa (or its Affiliates) in connection with the Kiniksa Development Activities or other activities performed or contemplated by Kiniksa under this Agreement for the Lead Antibody or Licensed Products, including CRO agreements, clinical trial agreements, institutional review board agreements, manufacturing agreements, supply agreements, laboratory services agreements, and other service provider agreements ("**Subsequent Clinical Study-Related Contracts**"). Following the completion of any applicable Kiniksa Transition Activities to which such Subsequent Clinical Study-Related Contracts may relate (or such earlier date as the Parties may agree), Kiniksa shall cooperate with and assist GNE, at GNE's reasonable request, in coordinating the transition of such Third Party subcontractor relationships from Kiniksa to GNE (including providing relevant contact information and making introductions to key personnel of such Third Parties or, if agreed by the Parties and permissible under the terms of the applicable subcontractor agreement, assigning such agreement to GNE).

7.4.2 Clinical Materials

To the extent permissible under the terms of any applicable patient consent form, the Materials transferred to GNE pursuant to the Tech Transfer shall include all available human biological patient samples Controlled by Kiniksa that relate to the development, manufacture or commercialization of the Lead Antibody or Licensed Product. As of the Execution Date, Kiniksa is not aware of any limitation in the applicable patient consent forms that would preclude it from transferring biological patient samples to GNE.

7.4.3 Ongoing Supporting Studies

For ongoing ancillary studies (chronic tox, stability, etc.) supporting the In Progress PN Study or necessary to maintain the IND for the Lead Antibody or applicable Licensed Product, following completion of the In Progress PN Study, upon GNE's request, Kiniksa will transfer any such study to GNE (or, as requested by GNE, enter into an arrangement that delegates the roles, rights, and responsibilities for such study between GNE, Kiniksa and the Third Party subcontractor supporting such study, including using reasonable efforts to amend the agreements between Kiniksa and such Third Party subcontractor for such study) and all data and information relating to such studies.

8. Regulatory

8.1 *Responsibility*

From the Effective Date until completion of the regulatory transfer of all US INDs for the Lead Antibody in accordance with Section 8.2 (Regulatory Transfer), Kiniksa shall retain (or shall cause its Affiliates to retain) sponsorship of the existing IND for the In Progress PN Study and Kiniksa shall be solely responsible, at its own cost, for all regulatory affairs related to the conduct of the In Progress PN Study. Prior to any transition of regulatory responsibility to GNE, Kiniksa shall be responsible for pursuing, compiling and submitting all Regulatory Filing documentation, and for interacting with Regulatory Authorities, for the In Progress PN Study. Notwithstanding the foregoing, Kiniksa shall seek (or cause its Affiliate to seek) input from GNE in preparing such regulatory materials and interactions, and GNE, at its own discretion, will have the right to (a) review and approve all written correspondence in advance of submission and (b)

approve scheduling of, and, to the extent permitted under Applicable Law, participate in all meetings with Regulatory Authorities.

After completion of the regulatory transfer in accordance with Section 8.2 (Regulatory Transfer) (including in the case that such transfer occurs prior to the completion of the In Progress PN Study), GNE will assume all regulatory responsibility with respect to the development of the Licensed Antibodies (including the Lead Antibody), and Licensed Products in the Field in the Territory. GNE shall be solely responsible, at its own cost, for all regulatory affairs related to Licensed Product in the Territory including the preparation and filing of INDs, BLAs, and other Regulatory Filings, as well as any or all governmental approvals required to develop, have developed, make, have made, use, have used, manufacture, have manufactured, import, have imported, sell and have sold Licensed Products. GNE shall be responsible for pursuing, compiling and submitting all Regulatory Filing documentation, and for interacting with regulatory agencies, for all Licensed Products in all countries in the Territory. GNE or its Affiliates shall own and file in their discretion all Regulatory Filings and Marketing Authorizations for all Licensed Products in all countries of the Territory. If the IND Transfer Request Date occurs prior to the completion of the In Progress PN Study, then after completion of the regulatory transfer for the In Progress PN Study in accordance with Section 8.2 (Regulatory Transfer), GNE will be solely responsible, at its own cost, for the conduct of such study and all regulatory affairs relating thereto.

8.2 *Regulatory Transfer*

In accordance with the applicable timelines and procedures set forth in the Tech Transfer Plan, Kiniksa shall provide to GNE copies of (a) the safety databases for the Lead Antibody, including all relevant historical clinical safety data, (b) all regulatory records and correspondence with the Regulatory Authorities that relate to the Lead Antibody, as listed in the Tech Transfer Plan, and (c) copies of the In Progress PN Study Data. Notwithstanding any provision to the contrary set forth in this Agreement, Kiniksa shall provide a final copy of the In Progress PN Study Data upon completion of the In Progress PN Study.

Promptly following the IND Transfer Request Date, Kiniksa shall (or shall cause its Affiliate(s) to) (i) transfer and assign to GNE all US INDs for the Lead Antibody, and (ii) provide to GNE all Regulatory Filings (other than ex-US INDs, except as set forth below) related to the Licensed Antibodies and Licensed Products, including clinical and safety databases, clinical study dossiers, regulatory correspondence, Regulatory Authority meeting minutes and study reports from completed non-clinical and clinical studies (including the In Progress PN Study Data). For all completed study reports, Kiniksa shall (or shall cause its Affiliate(s) to) provide to GNE documentation reasonably sufficient to confirm data reliability of the transferring regulatory materials, as required by Applicable Law, including original author signatures, raw data lists, and GLP and GCP compliance information. All documentation is to be provided in English.

Kiniksa shall be responsible for completing ex-US INDs for the In Progress PN Study and fulfilling all regulatory requirements related thereto, and following completion of the In Progress PN Study, shall close such ex-US INDs, *except* if the IND Transfer Request Date occurs prior to the completion of the In Progress PN Study, in which case GNE shall have the right, but not the obligation, to request transfer of ex-US IND sponsorship to GNE or its designee, and after such transfer GNE shall be solely responsible for all activities related to such ex-US INDs at its own cost, as further detailed in Section 8.1 (Responsibility).

8.3 *Right of Reference*
Kiniksa will (or will cause its Affiliates, as applicable, to) grant GNE a right of cross-reference to Kiniksa Regulatory Filings relating to the Licensed Antibody and Licensed Product to enable GNE to work on regulatory activities for the Licensed Antibodies and Licensed Products following the Effective Date and prior to Kiniksa's transfer and assignment of all INDs to GNE.

8.4 *Pharmacovigilance Agreement*
Within [***] days after the Effective Date, Parties shall execute a separate Pharmacovigilance Agreement specifying the procedures and timeframes for compliance with Applicable Law pertaining to safety reporting of the Licensed Antibody and Licensed Product(s) and their related activities, and appropriate audit rights.

9. Payment

9.1 *Initiation Payment*
Within thirty (30) days after the Effective Date and receipt of an invoice from Kiniksa therefor, GNE shall pay to Kiniksa eighty million US dollars (US$80,000,000).

9.2 *Fee upon Satisfaction of Clinical Supply by Required Supply Dates*
Subject to the following two sentences, if Kiniksa supplies (a) the Initial Drug Supply by no later than its Required Supply Date; and (b) the DP Resupply by no later than its Required Supply Date; then within thirty (30) days after GNE's receipt of an invoice from Kiniksa therefor (to be issued no earlier than the date that the DP Resupply is delivered and accepted by GNE in accordance with Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody)), GNE shall pay Kiniksa twenty million US dollars (US$20,000,000) (such amount being the **"Clinical Supply Fee"**). With respect to the Initial Drug Supply only, if Kiniksa fails to deliver the Initial Drug supply by its Required Supply Date, or if the Initial Drug Supply fails to comply with the delivery or release requirements of Section 6.1.2.5 (Additional Clinical Supply Obligations and Specifications), then (i) GNE will notify Kiniksa of such noncompliance, (ii) Kiniksa will have [***] days to complete delivery of, or cure any nonconformance with respect to the Initial Drug Supply, and (iii) if Kiniksa so completes the delivery or cures the nonconformance of the Initial Drug Supply within such [***] day period, then Kiniksa will have satisfied the Required Supply Date for the Initial Drug Supply for the purposes of this Section 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Dates). If there is a GNE Caused Required Date Failure for the Initial Drug Supply or the DP Resupply, then, to the extent the Clinical Supply Fee is not already payable by GNE under this Section 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Dates), GNE shall pay the Clinical Supply Fee within [***] days after the date that Kiniksa completes the delivery of both the Initial Drug Supply and the DP Resupply in accordance with Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody) and GNE's receipt of an invoice from Kiniksa therefor (to be issued no earlier than the date that both the Initial Drug Supply and the DP Resupply are delivered and accepted by GNE in accordance with Section 6.1.2 (Kiniksa Responsibility for Certain Clinical Supply of Lead Antibody)).

9.3 *Development and Regulatory Event Payments*
GNE shall pay (a) up to a total of [***] US dollars (US$[***]) with respect to the first achievement of each of the corresponding development milestone events set forth in Table 9.3 below for the first Licensed Product to achieve such event; and (b) subject to adjustment pursuant to Section

9.3.1 (APA Milestone Shortfall), up to a total of [***] US dollars (US$[***]) for the achievement of each development milestone event set forth in Table 9.3 below for each distinct Licensed Product to achieve such development milestone event after another Licensed Product has already achieved the same development milestone event.

9.3.1 APA Milestone Shortfall

If, for a given development milestone event listed in Table 9.3 below, Kiniksa is obligated to pay Biogen under the Biogen APA (as evidenced to GNE by disclosure of the applicable provision of the Biogen APA for such development milestone event on an unredacted basis) an amount greater than specified for such development milestone event in Column B of Table 9.3 (the difference in such amounts for a given development milestone event specified below, a "**APA Milestone Shortfall**"), then the development milestone payment in Column B of Table 9.3 payable upon achievement of such development milestone event shall be increased by the amount of such APA Milestone Shortfall; *provided* that in no event shall the amount payable by GNE for a given development milestone event under Column B of Table 9.3 exceed the amount specified in Column A of Table 9.3 for such development milestone event.

Table 9.3- Development and Regulatory Event Payments

Development Milestone Event	**Development Milestone Payment (US Dollars (in millions))**	
	Column A	**Column B**
	First Licensed Product	**Each Licensed Product other than the First Licensed Product**
[***]	[***]	[***]

9.3.2 Skipped Milestones

Each development milestone payment for a given Licensed Product shall be paid only once for a given Licensed Product, the first time such Licensed Product reaches such development event, regardless of the number of times such events are reached for such Licensed Product. For clarity, (i) the identity of the "first Indication", the "second Indication", and "third Indication" for a given Licensed Product shall be separately determined for each development milestone event based solely upon the order in which such particular milestone event is achieved with respect to the various Indications for such Licensed Product; and (ii) a given development milestone event shall be payable at the development milestone payment amount set forth in Column A of Table 9.3 only upon the first achievement of such development milestone event by a Licensed Product, and all Licensed Products to achieve a given development event after any other Licensed Product has already achieved the same such development milestone event shall be payable at the amount set forth in Column B of Table 9.3, subject to adjustment pursuant to Section 9.3.1 (APA Milestone Shortfall). If one or more development milestone events is not achieved for a given Licensed Product and given Indication prior to the achievement of a subsequent development milestone event for such Licensed Product in such Indication, then the development milestone payment(s) due for such skipped development milestone event(s) will be payable upon the first achievement of any subsequent development milestone event by such Licensed Product in such Indication, (*e.g.*, the occurrence of the development milestone event "[***]" will trigger the development milestone payments for "[***]" and "[***]," to the extent such development milestone payments have not already been paid for such Licensed Product in such

Indication). A development milestone event specific to one territory will not be deemed to be skipped solely because a subsequent development milestone event was achieved in a different territory.

9.3.3 Development Milestone Payment

Upon reaching a given development milestone event, GNE shall notify Kiniksa within [***] days of such achievement and the development milestone payment specified therefor shall be paid by GNE to Kiniksa within [***] days from GNE's receipt of an invoice from Kiniksa issued by Kiniksa after its receipt of such notice.

9.3.4 Development Milestone Achievement Disputes

If Kiniksa believes any such development milestone event under this Section 9.3 (Development and Regulatory Event Payments) has occurred and has not received a written notice of the same from GNE, then it shall so notify GNE in writing and shall provide to GNE documentation or other information that supports its belief. Any dispute under this Section 9.3 (Development and Regulatory Event Payments) that relates to whether a development milestone event has occurred that cannot be resolved among the Party's respective Alliance Managers within [***] days may be referred to both Party's executive officers pursuant to Section 20.2 (Disputes) by either Party, and, to the extent not resolved by the executive officers thereunder within [***] days, may be resolved in accordance with Section 20.3 (Jurisdiction; Consent to Forum).

9.4 *Sales Based Events*

On a Licensed Product-by-Licensed Product basis, GNE shall pay the following one-time payments to Kiniksa, up to a total of [***] US dollars (US$[***]), based on Calendar Year Net Sales on which royalties are paid for such Licensed Product in the Territory during the applicable Royalty Term.

Sales Milestone Event	**Payment US$ in Millions**
[***]	[***]
TOTAL	**US$**[***]

On a Licensed Product-by-Licensed Product basis, each of the sales-based event payments shall be paid no more than once during the applicable Royalty Term, within [***] days after the end of the Calendar Year in which the applicable sales threshold event first occurs for such Licensed Product in the Territory, and shall be non-refundable.

If Kiniksa believes any such sales threshold milestone event under this Section 9.4 (Sales Based Events) has occurred and has not received a written notice of same from GNE, it shall so notify GNE in writing and shall provide to GNE documentation or other information that supports its belief. Any dispute under this Section 9.4 (Sales Based Events) that relates to whether a sales threshold milestone event has occurred that cannot be resolved among the Party's respective Alliance Managers may be referred to both Party's executive officers pursuant to Section 20.2 (Disputes) by either Party, and, to the extent not resolved by the executive officers thereunder within [***] days, may be resolved in accordance with Section 20.3 (Jurisdiction; Consent to Forum).

9.5 *Royalty Payments*

9.5.1 *Royalty Term*
On a Licensed Product-by-Licensed Product basis, GNE shall pay to Kiniksa royalties on Net Sales of such Licensed Products during the Royalty Term at the rates set forth in Section 9.5.2 (Royalty Rates).

9.5.2 *Royalty Rates*
On a Licensed Product-by-Licensed Product basis, the following royalty rates shall apply to the respective tiers of aggregate Calendar Year Net Sales of a given Licensed Product in the Territory during the Royalty Term (as determined in the applicable country), on an incremental basis, as follows:

Tier of Calendar Year Net Sales for such Licensed Product (in Million US$)	**Percent (%) of Net Sales for such Licensed Product**
[***]	[***]

For example, if Net Sales of a given Licensed Product for a given Calendar Year during the Royalty Term for such Licensed Product (as determined in the applicable country), are US$[***], then royalties owed to Kiniksa on such Net Sales of such Licensed Product for that Calendar Year shall equal [***] US dollars (US$[***]) calculated as follows:

([***])+([***])+([***]) = US$[***] royalty payment

For the purpose of calculating royalties of a given Licensed Product, Calendar Year Net Sales and the royalty rates shall be subject to the following adjustments, as applicable:

9.5.3 *Combination Product*
If GNE or its Affiliates sells a Licensed Product as a Combination Product in any country in the Territory in any Calendar Quarter, then the Net Sales for such Combination Product on which royalties are payable shall be calculated by [***]

9.5.4 *No Valid Claim*
For a given Licensed Product, if in a given country within the Territory there is no Valid Claim that Covers such Licensed Product and no applicable Regulatory Exclusivity remains in such country for such Licensed Product, then the royalty payments due to Kiniksa for such Licensed Product in such country shall be reduced by [***] percent ([***]%) continuing until the last day of the applicable Royalty Term for such Licensed Product in such country.

9.5.5 Competing Drugs
Upon the first entry in a given country of a Competing Drug, the royalties in such country for such Licensed Product shall be reduced as follows: (a) if in any Calendar Quarter at any time after entry of a Competing Drug there has been a decline of the Sales of the applicable Licensed Product in such country greater than [***] of the level of the Sales of such Licensed Product achieved in [***] immediately prior to such entry, then the royalty payments due to Kiniksa for such Licensed Product in such country shall be reduced by [***]; and (b) if in any

Calendar Quarter at any time after entry of a Competing Drug there has been a decline of the Sales of the applicable Licensed Product in such country greater than [***] of the level of the sales of such Licensed Product achieved in [***] immediately prior to such entry, then the royalty payments due to Kiniksa for such Licensed Product in such country shall be reduced by [***]. For purposes of this Section 9.5.5 (Competing Drugs), (x) a "**Competing Drug**" means, with respect to a Licensed Product, a therapeutic product that is not produced, licensed or owned by the GNE Group and that (i) [***], (ii) [***], and (iii) [***]; and (y) "**Bioequivalent**" or **"Bioequivalence"** means, a biological product that (i) is highly similar to the Licensed Product notwithstanding minor differences in clinically inactive components; and (ii) has no clinically meaningful differences between the biological product and the Licensed Product in terms of the safety, purity, and potency.

9.5.6 Third Party Payments

Kiniksa shall remain responsible for and pay any payments owed to any Third Party pursuant to an arrangement in existence as of the Execution Date that relates to the Licensed IP, Licensed Antibodies, or Licensed Product, including all payment obligations and other contractual obligations under the Biogen APA and the Retained Contracts (as defined in the Biogen APA).

The amount of royalties payable to Kiniksa as specified in Section 9.5.2 (Royalty Rates) for any Licensed Product in any country shall be reduced by [***] of the amount of any royalty payments paid by GNE or any of its Affiliates to any Third Party in consideration for the license of Patent Rights in such country under any agreement entered into with any such Third Party after the Effective Date if, at the time of manufacture, use or sale of such Licensed Product, such Patent Rights would be infringed by the manufacture, use or sale of such Licensed Product in such country in the absence of such a license; *provided* that, in no event, shall the royalty payments as specified in Section 9.5.2 (Royalty Rates) with respect to such Licensed Product in a country be reduced by operation of this Section 9.5.6 (Third Party Payments) by more than [***] of what would otherwise be owed as specified in Section 9.5.2 (Royalty Rates); and [***].

9.5.7 Maximum Reductions

In no event shall the royalty paid to Kiniksa for Net Sales of Licensed Products hereunder be reduced from the applicable royalty rates set forth above by more than an amount equal to [***] of the royalties otherwise due for Net Sales of such Licensed Products, per the applicable royalty rates set forth above. [***].

9.5.8 Disposition of Rights to Payment

If Kiniksa intends to enter into any transaction with a Third Party for the sale, assignment, transfer or other disposition by Kiniksa of any rights to the payments due or payable by GNE to Kiniksa pursuant to this Agreement (a "**Disposition Transaction**"), then Kiniksa shall notify GNE of such intention in writing. At any time within [***] days after GNE receives such notice of the proposed Disposition Transaction from Kiniksa (the "**ROFN Exercise Period**"), GNE will have the option to notify Kiniksa (an "**ROFN Notice**") that it wishes to negotiate a buy-out by GNE of the rights to the payments due and payable by GNE to Kiniksa pursuant to this Agreement that Kiniksa proposes to sell, assign, transfer or otherwise dispose of in the Disposition Transaction (the "**Payment Rights**"). If GNE provides a ROFN Notice within the ROFN Exercise Period, Kiniksa shall negotiate exclusively and in good faith with GNE concerning the terms of a buy-out by GNE of the Payment Rights for a period of [***] after GNE delivers the ROFN Notice (the "**Negotiation Period**"). Kiniksa shall not negotiate or otherwise discuss a Disposition Transaction with any Third Party other than GNE, or provide any information or terms relating to a Disposition Transaction to any Third Party, until after the later

to expire of the ROFN Exercise Period and the Negotiation Period with respect to such Disposition Transaction. If GNE (a) notifies Kiniksa that it does not wish to negotiate a buy-out of the applicable Payment Rights; (b) fails to deliver a ROFN Notice to Kiniksa during the ROFN Exercise Period; or (c) delivers a ROFN Notice to Kiniksa during the ROFN Exercise Period but the Parties fail to reach agreement on the terms of the buy-out of the applicable Payment Rights or to execute a definitive agreement with respect to such buy-out prior to the expiration of the Negotiation Period, then in each case ((a), (b), or (c)), Kiniksa shall be free to negotiate and enter into the Disposition Transaction with respect to the same Payment Rights with any other party; *provided*, *however*, that in the case of clause (c), during the [***] period after GNE's last timely written offer (if any), Kiniksa may only enter into a Disposition Transaction that is on terms that, when taken as a whole, are more favorable to Kiniksa than the terms in GNE's last written offer.

9.6 *Disclosure of Payments*
Kiniksa acknowledges that GNE may be obligated to disclose this financial arrangement, including all fees, payments and transfers of value, as may be advisable or required under Applicable Law, including the US Sunshine Act.

10. Accounting and reporting

10.1 *Timing of Payments*
GNE shall calculate royalties on Net Sales quarterly as of March 31, June 30, September 30 and December 31 and shall pay royalties on Net Sales within [***] days after the end of each Calendar Quarter in which such Net Sales occur.

10.2 *Late Payment*
Any payment under this Agreement that is not paid on or before the date such payment is due shall bear interest at [***] from the due date until paid in full or, if less, the maximum interest rate permitted by Applicable Law.

10.3 *Method of Payment*
Royalties on Net Sales and all other amounts payable by GNE hereunder shall be paid by GNE in US Dollars (the "**Payment Currency**") to account(s) designated by Kiniksa.

10.4 *No Refunds*
Except as expressly provided under Section 9.5.6 (Third Party Payments), Section 9.5.7 (Maximum Reductions) and Section 12.3 (Over- or Underpayment), all payments under this Agreement will be irrevocable, non-refundable, and non-creditable; *provided* that with respect to the calculation of Net Sales for an applicable Calendar Quarter or Calendar Year, GNE may make credit or debit adjustments for royalty related payments made for such reporting period in a following period to reflect additional information that GNE's accounting department becomes aware of after such reporting period.

10.5 *Currency Conversion*
When calculating the Sales of any Licensed Product that occur in currencies other than the Payment Currency, GNE shall convert the amount of such sales into Swiss Francs and then into the Payment Currency using GNE's then-current internal foreign currency translation method actually used on a consistent basis in preparing its audited financial statements (at the Effective Date, YTD average rate as reported by Reuters).

10.6 *Blocked Currency*
In a given country, if by reason of Applicable Law (for example, governmental restrictions on foreign exchange trade) the local currency is blocked and cannot be removed from such country, GNE will notify Kiniksa in writing and

(a) Kiniksa will have the right to receive the applicable royalties of Net Sales in such country in local currency by deposit in a local bank designated by Kiniksa; or

(b) if such local currency payment is not allowed by reason of Applicable Law or if otherwise requested by Kiniksa, then the royalties related to such Net Sales in such country shall continue to be accrued and shall continue to be reported, but such royalties will not be paid until the sales proceeds related to such Net Sales may be removed from such country. At such time as GNE, its Affiliates or their Sublicensees, as the case may be, is able to remove the sales proceeds related to such Net Sales from such country, GNE shall also pay such accrued royalties in Payment Currency using the actual exchange rate that is used to remove such sales proceeds from such country.

10.7 *Reporting*
With each royalty payment paid pursuant to Section 10.1 (Timing of Payments) for an applicable Calendar Quarter GNE shall provide Kiniksa in writing for the relevant Calendar Quarter on a Licensed Product-by-Licensed Product basis the following information:

(a) Sales (in Swiss Francs);
(b) Net Sales in Swiss Francs;
(3) adjustments made pursuant to Section 9.5.3 (Combination Product);
(4) Net Sales in Swiss Francs after adjustments made pursuant to Section 9.5.3 (Combination Product);
(5) exchange rate used for the conversion of Net Sales from Swiss Francs to the Payment Currency pursuant to Section 10.5 (Currency Conversion);
(6) Net Sales after adjustments made pursuant to Section 9.5.3 (Combination Product) in the Payment Currency;
(7) royalty rate pursuant to Section 9.5.2 (Royalty Rates);
(8) adjustments made pursuant to Sections 9.5.4 (No Valid Claim) through Section 9.5.7 (Maximum Reductions); and
(9) total royalty payable in the Payment Currency after adjustments made pursuant to Sections 9.5.4 (No Valid Claim) through Section 9.5.7 (Maximum Reductions).

11. Taxes

Kiniksa shall pay all sales, turnover, income, revenue, value added, and other taxes levied on account of any payments accruing or made to Kiniksa under this Agreement.

If provision is made in law or regulation of any country for withholding of taxes of any type, levies or other charges with respect to any royalty or other amounts payable under this Agreement to Kiniksa, then GNE shall promptly pay such tax, levy or charge for and on behalf of Kiniksa to the proper Governmental Authority, and shall promptly furnish Kiniksa with receipt of payment. GNE shall be entitled to deduct any such tax, levy or charge actually paid from royalty or other payment due Kiniksa or be promptly reimbursed by Kiniksa if no further payments are due to Kiniksa. Each Party agrees to reasonably assist the other Party in claiming exemption from such deductions or withholdings under double taxation or similar agreement or treaty from time to time in force and in minimizing the amount required to be so withheld or deducted.

12. Auditing

12.1 *Kiniksa Right to Audit*
GNE shall keep, and shall require its Affiliates and Sublicensees to keep, full, true and accurate books of account containing all particulars that may be necessary for the purpose of calculating all royalties payable under this Agreement and accurate records of Sales and Net Sales by GNE and its Affiliates and Sublicensees in sufficient detail to allow payments due hereunder to be determined accurately. Such books of accounts shall be kept at their principal place of business. At the expense of Kiniksa, Kiniksa shall have the right to engage an internationally recognized independent public accountant reasonably acceptable to GNE to perform, on behalf of Kiniksa, an audit of such books and records of GNE and its Affiliates that are deemed necessary by the independent public accountant to report on Net Sales of Licensed Product for the period or periods requested by Kiniksa and the correctness of any financial report or payments made under this Agreement.

Upon timely request and at least [***] prior written notice from Kiniksa, such audit shall be conducted for those countries Kiniksa has specifically requested, during regular business hours, at such place or places where such records are customarily kept by GNE or its Affiliate, in such a manner as to not unnecessarily interfere with GNE's normal business activities. Such audit shall be limited to results in the [***] prior to audit notification, and [***]. If Kiniksa does not request an audit of a given Calendar Year [***], then Kiniksa will be deemed to have accepted the royalty payments and reports in such Calendar Year.

Such audit shall not be performed more frequently than [***] nor more frequently than [***]. All information, data documents and abstracts herein referred to shall be used only for the purpose of verifying Sales and Net Sales amounts, shall be treated as GNE's Confidential Information subject to the obligations of this Agreement and need neither be retained more than [***] after completion of an audit hereof, if an audit has been requested; nor more than [***] from the end of the Calendar Year to which each shall pertain; nor more than [***] after the date of termination of this Agreement.

12.2 *Audit Reports*
The auditors shall only state factual findings in the audit reports and shall not interpret this Agreement. The auditors shall share all draft audit findings with GNE before sharing such findings with Kiniksa and before the final audit report is issued. The final audit report shall be shared with GNE at the same time it is shared with Kiniksa.

12.3 *Over-or Underpayment*
If the audit reveals an overpayment, then GNE may, from future payments owed by GNE, deduct the amount of the overpayment or, if no further royalty payments are owed by GNE, Kiniksa shall reimburse GNE for the amount of the overpayment within [***] days. If the audit reveals an underpayment, GNE shall make up such underpayment with the next royalty payment (but in any event no later than [***] days after GNE's receipt of the report so concluding) or, if no further royalty payments are owed by GNE, GNE shall pay Kiniksa for the amount of the underpayment within [***] days after receipt of the final audit report. GNE shall pay for the audit costs if the underpayment of GNE exceeds [***] percent ([***]%) of the aggregate amount of royalty payments payable in any Calendar Year subject to the audit. Section 10.2 (Late Payment) shall apply to payments due pursuant to this Section 12.3 (Over-or Underpayment).

13. Intellectual Property

13.1 *Ownership of Inventions*
Kiniksa shall own all Kiniksa Inventions, GNE shall own all GNE Inventions, and Kiniksa and GNE shall jointly own all Joint Inventions. Kiniksa and GNE each shall require all of its, and to the extent conducting activities under this Agreement, their respective Affiliates' and Sublicensees', employees to assign all Inventions made by them to GNE and Kiniksa, as the case may be. The determination of inventorship for Inventions shall be in accordance with US inventorship laws as if such Inventions were made in the US.

Subject to the licenses granted to GNE under Section 2.1 (Licenses), Kiniksa and GNE will each have an equal undivided share in the Joint Patent Rights, without obligation to account to the other for exploitation thereof, or to seek consent of the other Party for the grant of any license thereunder.

With respect to any other Know-How (other than Kiniksa Inventions, GNE Inventions, and Joint Inventions) arising from the Parties' activities under this Agreement, (a) Kiniksa shall own such Know-How made by employees of Kiniksa solely or jointly with a Third Party, (b) GNE shall own all Know-How made by employees of the GNE Group solely or jointly with a Third Party, and (c) the Parties shall jointly own the Joint Know-How.

If there is a disagreement between the Parties with respect to the inventorship of Inventions, then the Parties shall jointly select a reputable patent attorney registered before the United States Patent and Trademark Office with over ten (10) years of experience in the relevant subject matter and submit such disagreement to the mutually-selected patent attorney for resolution under United States patent law. The decision of such patent attorney with respect to inventorship shall be final, unchallengeable, and bind both Parties. The Parties shall share equally the expenses of such patent attorney for such inventorship determination.

13.2 *Prosecution of Licensed Patent Rights*
As between the Parties, GNE shall, at its own expense and discretion, Handle all Licensed Patent Rights solely owned by Kiniksa. At GNE's expense and reasonable request, Kiniksa shall cooperate, in all reasonable ways, with the Handling of all such Licensed Patent Rights. GNE will keep Kiniksa reasonably informed of substantive matters relating to prosecution of Licensed Patent Rights. If GNE decides that it does not desire to Handle (including abandon) any such Licensed Patent Right, then it shall promptly advise Kiniksa thereof. Notwithstanding any provision to the contrary set forth in this Agreement, GNE shall have no right to Handle the Patent Rights within the Biogen Background IP Rights.

13.3 *Prosecution of Other Patent Rights*
In addition to Section 13.2 (Prosecution of Licensed Patent Rights), GNE shall, at its own expense and discretion, Handle (including abandon) all other Patent Rights that contain one or more claims that Cover any Licensed Antibody or Licensed Product, including all Joint Patent Rights.

13.4 *Abandonment of Handling*

13.4.1 Non-Strategic Abandonment
If GNE decides to abandon or allow to lapse, or otherwise determines not to Handle a Patent Right in accordance with Section 13.2 (Prosecution of Licensed Patent Rights) or Section 13.3

(Prosecution of Other Patent Rights) in any country or region in the Territory (including failing to timely respond to a patent office communication without timely filing a continuation or divisional application to preserve such Patent Right), then GNE will provide prior written notice to Kiniksa of such intention so as to provide Kiniksa with a reasonable amount of time to meet any applicable deadline to preserve such Patent Rights in such country or region. In such case, upon written notice to GNE, Kiniksa will have the right (but not the obligation) to assume responsibility for Handling such Patent Right in such country or region at its sole expense, and GNE will promptly deliver to Kiniksa copies of all relevant documents necessary to Handle such Patent right in such country or region.

13.4.2 Abandonment for Strategic Rationale

Notwithstanding Section 13.4.1 (Non-Strategic Abandonment), if such Patent Right is not the sole Patent Right that Covers the Lead Antibody within such Licensed Patent Rights or Joint Patent Rights in such country or region and GNE provides prior written notice to Kiniksa of proposed abandonment of such Patent Rights based on a reasonable strategic rationale, GNE shall concurrently provide Kiniksa with the basis of such strategic rationale, and Kiniksa shall have no right to assume responsibility for Handling such Patent Right; *provided, however*, if there is a disagreement between the Parties with respect to the reasonableness of the strategic rationale for abandonment, then the Parties shall jointly select a reputable patent attorney registered before the United States Patent and Trademark Office with over ten (10) years of experience in the relevant subject matter and submit such disagreement to the mutually-selected patent attorney for his or her opinion as to the reasonableness of the strategic rationale for such abandonment. The decision of such patent attorney with respect to the reasonableness of such strategic rationale shall be final, unchallengeable, and bind both parties. The Parties shall share equally the expenses of such patent attorney for such decision.

13.5 *CREATE Act*

It is the intention of the Parties that this Agreement is a “joint research agreement” as that phrase is defined in 35 USC § 102(c) (AIA). In the event that either Party to this Agreement intends to overcome a rejection of a claimed invention covered by Joint Patent Rights or Licensed Patent Rights pursuant to the provisions of 35 USC §§ 102(a)-(d), such Party shall first obtain the prior written consent of the other Party. Following receipt of such written consent, such Party shall limit any amendment to the specification or statement to the patent office with respect to this Agreement to that which is strictly required by the applicable subsection of 35 USC § 102 and the rules and regulations promulgated thereunder and that is consistent with the terms and conditions of this Agreement (including the scope of the Kiniksa Development Activities and other Kiniksa Transition Activities). To the extent that the Parties agree that, in order to overcome a rejection of a claimed invention covered by Joint Patent Rights or Licensed Patent Rights pursuant to the provisions of the applicable subsection of 35 USC § 102, the filing of a terminal disclaimer is required or advisable, the Parties shall first agree on terms and conditions under which the patent application subject to such terminal disclaimer and the patent or application over which such application is disclaimed shall be jointly enforced, to the extent that the Parties have not previously agreed to such terms and conditions. In the event that GNE enters into an agreement with a Third Party with respect to the further research, development or commercialization of a Licensed Product, Kiniksa shall, upon GNE’s request, similarly enter into such agreement with such Third Party for the purposes of furthering the Parties’ objectives under this Agreement; *provided* that such agreement does not place any material obligation on Kiniksa.

13.6 *Infringement*

13.6.1 Notice

If either Party becomes aware of (a) any suspected infringement or misappropriation of any Licensed IP, Joint Patent Rights or Joint Know-How; or (b) the submission by any Third Party of an abbreviated BLA under the Biologics Price Competition and Innovation Act for any Licensed Product (each, an **"Infringement"**), that Party shall promptly notify the other Party and provide it with all details of such Infringement of which it is aware.

13.6.2 GNE Right

Within [***] days after GNE provides or receives a written notice as set forth in Section 13.6.1 (Notice) ("**Decision Period**"), GNE, in its sole discretion, shall decide whether or not to initiate a suit or action in the Territory regarding such Infringement involving any Licensed IP solely owned by Kiniksa or any Joint Patent Right or Joint Know-How and shall notify Kiniksa of its decision in writing ("**Suit Notice**"); *provided* that, if GNE declines to initiate such a suit or action, then it shall provide a Suit Notice to Kiniksa with sufficient time such that such Infringement Response will not be prejudiced.

If GNE decides to bring such a suit or take action, once GNE provides Suit Notice (an "**Infringement Response**"), GNE may immediately commence such suit or take such an action and Kiniksa shall provide all reasonable cooperation to GNE in connection with such Infringement Response. With respect to any Infringement Response relating to the Licensed Patent Rights solely owned by Kiniksa or the Joint Patent Rights, (a) GNE shall keep Kiniksa informed about such Infringement Response; (b) GNE shall not take any position with respect to, or compromise or settle, any such Infringement action in any way without first providing Kiniksa with (i) notice of GNE's preferred course of action and (ii) an opportunity to provide comments, which comments Kiniksa will provide promptly and in any event within [***] days from receipt of such notice from GNE and which comments GNE will consider in good faith; and (c) if GNE ceases to diligently pursue an Infringement Response, it shall inform Kiniksa in such a manner that such Infringement Response will not be prejudiced and Section 13.6.3 (Kiniksa Right) shall apply. All costs, including attorneys' fees, relating to such Infringement Response shall be borne solely by GNE.

13.6.3 Kiniksa Right

If (a) GNE informs Kiniksa that it does not intend to pursue any Infringement Response with respect to any Licensed Patent Right or Joint Patent Right, (b) within [***] days after the receipt of notice of any such Infringement, GNE has not commenced to take any Infringement Response with respect thereto, or (c) if GNE ceases to reasonably pursue any such Infringement Response, then in each case ((a) through (c)), Kiniksa shall thereafter have the right to commence suit or take action in the Territory against such Infringement and shall provide written notice to GNE of any such suit commenced or action taken by Kiniksa. With respect to any such suit or action commenced by Kiniksa, (i) Kiniksa shall keep GNE informed about such suit or action, and (ii) Kiniksa shall not take any position with respect to, or compromise or settle, any such Infringement action in any way without first providing GNE with (x) notice of Kiniksa's preferred course of action and (y) an opportunity to provide comments, which comments GNE will provide promptly and in any event within [***] days from receipt of such notice from Kiniksa and which comments Kiniksa will consider in good faith. All costs, including attorneys' fees, relating to such Infringement Response shall be borne solely by Kiniksa.

13.6.4 Enforcement Costs and Allocation of Recovery

The Party bringing suit or taking action ("**Initiating Party**") shall, except as provided below, pay all expenses of the suit or action, including the Initiating Party's attorneys' fees and court costs. Unless otherwise agreed by the Parties, and subject to the Parties' respective obligations under Article 15 (Indemnification), all monies recovered upon the final judgment or settlement of any action described in this Section 13.6 (Infringement) shall be used as follows:

(a) First, to reimburse the Parties for their reasonable out-of-pocket expenses in making such recovery (which amounts shall be allocated *pro rata* if insufficient to cover the totality of such expenses); and

(b) Second,

(i) [***]; and

(ii) [***].

13.6.5 Cooperation

If the Initiating Party believes it is reasonably necessary or desirable to obtain an effective remedy, upon written request the other Party agrees to be joined as a party to the suit or action, but shall be under no obligation to participate except to the extent that such participation is required as the result of its being a named party to the suit or action. At the Initiating Party's written request, the other Party shall offer reasonable assistance to the Initiating Party in connection therewith, and the Initiating Party will reimburse the other Party's reasonable out-of-pocket expenses (including attorneys' fees) incurred in rendering such assistance. The other Party shall have the right to participate and be represented in any such suit or action by its own counsel at its own expense.

13.6.6 Settlement

The Initiating Party may settle, consent judgment or otherwise voluntarily dispose of the suit or action ("**Settlement**") without the written consent of the other Party if such Settlement can be achieved without adversely affecting the other Party (including any of its Patent Rights). If a Settlement could adversely affect the other Party, then the written consent of the other Party will be required, which consent shall not be unreasonably withheld.

For any patent that is not a Licensed Patent Right or Joint Patent Right, GNE, in its sole discretion, shall decide whether or not to initiate such suit or action in the Territory. GNE shall have full discretion as to how it wishes to handle such suit and may reach Settlement and retain all damages, settlement fees or other consideration under any terms and conditions it desires and retain whatever. Only if a Settlement could adversely affect Kiniksa shall the written consent of Kiniksa be required, which consent shall not be unreasonably withheld.

13.7 *Defense*

If an action for infringement is commenced against either Party, its licensees or its sublicensees related to the discovery, development, manufacture, use or sale of a Licensed Product, then the Party against whom such action is brought shall have the right (but not the obligation) to defend such action, at its own expense. The other Party shall assist and cooperate with the defending Party, at the defending Party's expense, to the extent necessary in the defense of such suit. The defending Party shall have the right to settle the suit or consent to an adverse judgment thereto, in its sole discretion, unless such settlement or adverse judgment affects the rights or interests of the other Party or its Affiliates (including with respect to any Patent Rights Controlled by any of them), in which case such settlement or consent shall be subject to other Party's prior written consent, not to be unreasonably withheld. Subject to Section 15 (Indemnification), the defending

Party shall assume full responsibility for the payment of any award for damages, or any amount due pursuant to any settlement entered into by it with such Third Party.

13.8 *Common Interest Disclosures*
With regard to any information, opinions, or materials disclosed pursuant to this Agreement by one Party to each other regarding intellectual property or technology owned by Third Parties, the Parties agree that they have a common legal interest in determining whether, and to what extent, Third Party intellectual property rights may affect Licensed Antibodies or Licensed Products, and have a further common legal interest in defending against any actual or prospective Third Party claims based on allegations of misuse or infringement of intellectual property rights relating to Licensed Antibodies or Licensed Products. Accordingly, the Parties agree that all such information and materials obtained by Kiniksa and GNE from each other will be used solely for purposes of the Parties' common legal interests with respect to the conduct of this Agreement. All information, opinions, and materials will be treated as protected by the attorney-client privilege, the work product privilege, and any other privilege or immunity that may otherwise be applicable. By sharing any such information, opinions, and materials, neither Party intends to waive or limit any privilege or immunity that may apply to the shared information and materials. Neither Party shall have the authority to waive any privilege or immunity on behalf of the other Party without such other Party's prior written consent, nor shall the waiver of privilege or immunity resulting from the conduct of one Party be deemed to apply against any other Party. Notwithstanding the foregoing, neither Party's attorney represents the other Party.

13.9 *Patent Term Extensions*
GNE shall use reasonable efforts to obtain all patent term extensions or supplemental protection certificates or their equivalents in any country where applicable to the Licensed Patent Rights. Kiniksa shall cooperate with GNE with respect to such matters, including by timely conferring with GNE to ensure compliance with applicable filing deadlines, and conferring with GNE on the procedures to be followed by GNE to ensure such compliance. Kiniksa shall execute such authorizations and other documents and take such other actions as may be reasonably requested by GNE to obtain such patent term extensions or supplemental protection certificates or their equivalents, including designating GNE as its agent for such purpose as provided in 35 USC § 156. All filings for such patent term extensions or supplemental protection certificates or their equivalents shall be made by GNE.

14. Representations, Warranties and Covenants

14.1 *Representation and Warranties of Kiniksa*
Kiniksa represents and warrants to GNE that, as of the Execution Date:

14.1.1 Safety Data
Kiniksa has disclosed to GNE (i) the results of all preclinical testing and human clinical testing of Licensed Product generated by or on behalf of Kiniksa or its Affiliates, and (ii) all information in its Control concerning side effects, injury, toxicity or sensitivity reaction and incidents or severity thereof with respect to Licensed Product, except, in each case ((i) and (ii)), to the extent not yet made available to Kiniksa (or its Affiliates) with respect to the In Progress PN Study.

14.1.2 No Back-Up Licensed Antibodies
Except as set forth in Appendix 14.1.2, neither Kiniksa nor any of its Affiliates (a) is researching or developing any Licensed Antibody other than the Lead Antibody; or (b) possess any other Licensed Antibodies.

14.1.3 Third Party Patent Rights

To Kiniksa's Knowledge, except as disclosed to GNE prior to the Execution Date, there is no valid issued Patent Right owned by any Third Party that that would prevent GNE from making, having made, using, offering for sale, selling or importing Licensed Antibody or Licensed Product in the Territory as contemplated by this Agreement.

14.1.4 Licensed IP

(a) Appendix 14.1.4(a) contains a complete and accurate list of all patents and patent applications that Kiniksa owns that claim or disclose the Licensed Know-How; (b) Appendix 14.1.4(b) specifies the Existing Lead Antibody Patent Rights; (c) with respect to any Licensed IP Controlled, but not owned by Kiniksa, Appendix 14.1.4(c) specifies each Third Party agreement pursuant to which Kiniksa Controls such Licensed IP and contains a list of the Licensed Patent Rights Controlled by Kiniksa pursuant to each applicable Third Party agreement, which list is a complete and accurate list of the Licensed Patent Rights that were specifically identified in a table scheduling Patent Rights in each such applicable agreement as of the date of execution of the applicable agreement; and (d) Appendix 14.1.4(d) specifies any agreement with a Third Party pursuant to which Kiniksa has any payment obligations with respect to Licensed IP or the research, development, registration, use, manufacture, import, export, marketing, distribution, sale or use of Licensed Antibodies or Licensed Products in the Field in the Territory. Except as identified in Appendix 14.1.4 (c) or (d), (i) Kiniksa is the exclusive owner of all rights, title and interests in, or is the exclusive licensee of, the Licensed IP; and (ii) Kiniksa does not owe any financial obligations to any Third Party with respect to the Licensed IP or the research, development, registration, use, manufacture, import, export, marketing, distribution, sale or use of Licensed Antibodies or Licensed Products in the Field in the Territory.

14.1.5 Inventors

Kiniksa has obtained the assignment of, or a license under, all interest and all rights or licenses with respect to the Licensed Patent Rights as necessary to grant the licenses granted to GNE hereunder. All of Kiniksa's (and each of its Affiliates', contractors' and (sub)licensees') employees, officers and consultants have executed agreements requiring assignment to Kiniksa or its Affiliates of all Inventions and Know-How made by such individuals during the course of and as a result of their association with Kiniksa or its Affiliate.

14.1.6 Grants

To Kiniksa's Knowledge, Kiniksa has the lawful right (including any necessary rights from its Affiliates) to grant GNE and its Affiliates all of the rights and licenses described in this Agreement.

14.1.7 Authorization

Kiniksa represents and warrants to GNE that the execution, delivery and performance of this Agreement and the Biogen Side Letter by it and all instruments and documents to be delivered by it hereunder: (i) are within its corporate power; (ii) have been duly authorized by all necessary or proper corporate action; (iii) are not in contravention of any provision of its certificate of formation or limited liability company agreement; (iv) to its Knowledge, will not violate any law or regulation or any order or decree of any Governmental Authority; (v) will not violate the terms of any indenture, mortgage, deed of trust, lease, agreement, or other instrument to which it is a party or by which it or any of its property is bound, which violation would have an adverse effect on its financial condition or on its ability to perform its obligations hereunder; and (vi) do not require any filing or registration with, or the consent or approval of, any Governmental Authority

or any other person, which has not been made or obtained previously (other than approvals required under Antitrust Laws, Marketing Authorizations required for the sale of Licensed Products and filings with Regulatory Authorities required in connection with Licensed Products). Further, Kiniksa represents and warrants to GNE that there are no claims or investigations pending or threatened against Kiniksa or any of its Affiliates, at law or in equity, or before or by any Governmental Authority relating to the matters contemplated under this Agreement or the Biogen Side Letter that would materially adversely affect Kiniksa's ability to perform its obligations hereunder or thereunder, and neither Kiniksa nor any of its Affiliates is or will be under any obligation to any person, contractual or otherwise, that is conflicting with the terms of this Agreement or the Biogen Side Letter, or that would impede the fulfillment of Kiniksa's obligations hereunder or thereunder.

14.1.8 Control of Licensed IP

Except solely with respect to the In Progress PN Study Data or any other Know-How Controlled by Kiniksa Parent that is necessary for the completion of the In Progress PN Study by Kiniksa Parent in accordance with this Agreement (the **"PN Study Know-How"**), no Affiliate of Kiniksa Controls or otherwise holds rights to any Know-How or Patent Rights that, if Controlled by Kiniksa, would constitute Licensed Know-How or Licensed Patent Rights, or, with respect to Kiniksa's interest therein, Joint Know-How or Joint Patent Rights, respectively. If any such Know-How or Patent Right comes into the Control of any Affiliate of Kiniksa, then Kiniksa will promptly cause such Affiliate to transfer and assign such Know-How or Patent Right to Kiniksa and, upon such transfer and assignment, such Know-How or Patent Right will be included under the Licensed Know-How, Licensed Patent Rights, Joint Know-How, or Joint Patent Rights (as applicable) for purposes of this Agreement.

14.1.9 Control of In Progress PN Study Data

Kiniksa or its Affiliates own or Control all data and information generated by, or on behalf of, Kiniksa, in the conduct of the In Progress PN Study.

14.1.10Validity of Patent Rights

To Kiniksa's Knowledge, (a) there are no inventorship disputes concerning any Existing Licensed Patent Rights, and (b) such Existing Licensed Patent Rights are (i) being diligently Handled where filed within the Territory and (ii) valid and enforceable.

14.1.11Ownership and Legitimacy of Licensed Know-How, Kiniksa Cell Line Materials and Inventory

To Kiniksa's Knowledge, the Licensed Know-How existing as of the Effective Date (a) is legitimately in the Control of Kiniksa and has not been misappropriated from any Third Party (or Kiniksa Affiliate); and (b) Kiniksa has taken reasonable measures to protect the confidentiality of such Know-How.

Kiniksa or its Affiliate is the sole and exclusive owner of, or otherwise has all rights necessary to transfer and assign or license to GNE, as applicable, in accordance with this Agreement with respect to, all existing clinical and non-clinical grade drug product and drug substance, active pharmaceutical ingredient, intermediates and raw materials associated with Licensed Antibodies, as well as any other existing Materials (such as reference standards and retention samples, critical assay reagents and cell banks), and packaging associated with such Licensed Antibodies and Licensed Products contained therein, and the Kiniksa Cell Line Materials, in each case, free and clear of all liens and all such Inventory and Kiniksa Cell Line Materials have, to the extent owned by Kiniksa, been developed and produced in compliance in all material respects with all GMP and Applicable Laws. To the Knowledge of Kiniksa, there are no material

defects in any of the Inventory or Kiniksa Cell Line Materials. The Kiniksa Cell Line Materials are suitable to be used as starting material for (i) further manufacturing of the Lead Antibody and (ii) the production of a working cell bank. To Kiniksa's Knowledge, all existing Inventory and Kiniksa Cell Line Materials are accurately listed in the Tech Transfer Plan.

14.1.12Existing Clinical Supply of Lead Product

(a) Kiniksa has the quantity of drug product, drug substance and placebo required for the Initial Drug Supply and DP Resupply as set forth in Appendix 6.1.2.2; (b) Kiniksa has sufficient additional quantities of drug product and placebo to conduct the In Progress PN Study; (c) Kiniksa or its Affiliate is the sole and exclusive owner of all such drug product, drug substance and placebo free and clear of all liens; and (d) all such drug product, drug substance and placebo satisfy the specifications and quality requirements for such drug substance, drug product and placebo as in existence as of the Execution Date and have been produced in compliance in all material respects with all GMP and Applicable Laws. To the Knowledge of Kiniksa, there are no defects in any such drug product, drug substance and placebo.

14.1.13Existing Clinical Studies

There are no Clinical Studies or other development activities being conducted by or on behalf of Kiniksa (or any of its Affiliates) relating to Licensed Antibodies or Licensed Products other than the In Progress PN Study; and all Clinical Studies conducted by or on behalf of Kiniksa (or any of its Affiliates) relating to Licensed Antibodies or Licensed Products have been conducted in accordance with Applicable Law, including good laboratory practices, good clinical practices, good manufacturing practices (and each of their foreign equivalents), and applicable privacy, data protection and informed consent requirements.

14.1.14DP Resupply

Kiniksa reserved [***] drug product sterile fill-finish manufacturing slots with its contract manufacturing organization sufficient to enable a delivery of the DP Resupply on or before [***], and in any event by no later than [***].

14.2 *Representation, Warranties and Covenants as to Biogen APA*

14.2.1 Biogen APA

Kiniksa represents, warrants and covenants to GNE as of the Effective Date that (a) Kiniksa Parent has properly assigned, and Kiniksa has properly assumed, the Biogen APA in its entirety (including all of Kiniksa Parent's obligations and rights under the Biogen APA and all of Kiniksa Parent's (and any of its Affiliates) assets to which the Biogen APA relates (including the Licensed IP transferred to Kiniksa under the Biogen APA)), except for the PN Study Know-How, and such assignment and assumption of the Biogen APA was consummated in conformance with all requirements therefor under the Biogen APA; (b) Kiniksa has provided GNE with a true, correct and complete copy of the Biogen APA (redacted only for non-material obligations, Kiniksa's financial obligations under the Biogen APA, or other obligations that, in each case, do not impact GNE's obligations to Kiniksa, or GNE's rights, under this Agreement); (c) subject only to the terms of the Biogen Side Letter and the Biogen APA Amendment, the Biogen APA remains in full force and effect; (d) neither Kiniksa nor Biogen has provided any notice or other communication to the other party thereunder of any default, breach, potential default or potential breach, in each case, under the Biogen APA; and (e) to Kiniksa's Knowledge, no event has occurred or circumstance exists that (with or without notice or lapse of time) would give either Biogen or Kiniksa the right to declare a breach or terminate the Biogen APA. With respect to any PN Study Know-How, Kiniksa covenants that it will cause Kiniksa Parent to transfer and

assign such Know-How to Kiniksa promptly following the earlier of completion of the In Progress PN Study or the IND Transfer Request Date, as set forth in, and in accordance with, the Tech Transfer Plan and Section 8.2 (Regulatory Transfer), and, upon such transfer and assignment, such Know-How will be included under the Licensed Know-How for purposes of this Agreement.

14.2.2 Maintenance of Biogen APA; Breach Notices and Other Material Correspondence

During the Agreement Term, Kiniksa shall (a) satisfy all of its material obligations to Biogen (including making all payment obligations) under the Biogen APA and the Biogen Side Letter; (b) not commit any act or omission that would cause a breach of or give rise to a right of Biogen to terminate the Biogen APA and take all steps in its control to maintain in full force and effect, the Biogen APA and the Biogen Side Letter; (c) not assign or otherwise transfer, in whole or in part, to any party (except an assignment to a party to whom this Agreement is assigned as permitted under Section 20.4 (Assignment)) the Biogen APA or the Biogen Side Letter; (d) not amend, restate, amend and restate, or otherwise modify the Biogen APA or the Biogen Side Letter in a manner that would impact or otherwise limit the rights or expand the obligations of GNE under this Agreement, without the prior written consent of GNE; (e) not terminate in whole or in part the Biogen APA or the Biogen Side Letter or otherwise exercise or waive any rights it may have under the Biogen APA or the Biogen Side Letter in a manner that would impact or otherwise limit the rights or expand the obligations of GNE with respect to the Licensed IP, the Joint Patent Rights or the Joint Know-How under this Agreement, without the prior consent of GNE; (f) provide GNE with prompt written notice of any notice or other communication of any claim of a breach or default under, or termination of, the Biogen APA or the Biogen Side Letter made by either Kiniksa or Biogen (or any party acting on behalf of either such party) whether under Article 8 of the Biogen APA or otherwise; and (g) promptly send to GNE copies of all other material correspondence to or from Biogen (or an party acting on either such party's behalf) that relates to a potential or actual breach or default of the Biogen APA that could give rise to a right for Biogen to terminate the Biogen APA or other material correspondence relating to a Biogen claim with respect to Biogen's rights, or Kiniksa's obligations, as to or related to any of the Licensed IP, the Joint Patent Rights, or the Joint Know-How. The notices provided to GNE under the foregoing clause (f) and (g) shall include in reasonable detail the substance and circumstances of such claims and Kiniksa's planned response and other potential actions to ensure that the Biogen APA remains in full force and effect. In addition, Kiniksa shall, at the request of GNE, promptly have its appropriate representatives and its legal counsel meet with GNE and its legal counsel to discuss such claims and Kiniksa's planned response and other potential actions to ensure that the Biogen APA remains in full force and effect.

14.2.3 GNE Opportunity to Cure APA Breach Claims on Kiniksa's Behalf

If, during the Agreement Term, Biogen (or any other party acting on Biogen's behalf) (a) makes any claim of a breach or default of the Biogen APA by Kiniksa (or by any party acting on Kiniksa's behalf, including GNE), or (b) seeks to terminate the Biogen APA, in whole or in part, (collectively (a) – (b), an "**APA Breach Claim**"), then to the extent Kiniksa has not yet done so under Section 14.2.2 (Maintenance of Biogen APA; Breach Notices and Other Material Correspondence), Kiniksa shall promptly notify GNE of such APA Breach Claim including in reasonable detail the substance and circumstances of such APA Breach Claim and Kiniksa's planned response and other potential actions to ensure that the Biogen APA remains in full force and effect. In addition, Kiniksa shall, at the request of GNE, promptly have its appropriate representatives and its legal counsel meet with GNE and its legal counsel to discuss such claims and Kiniksa planned response and other actions to ensure that the Biogen APA remains in full force and effect.

In addition, at the request of GNE, Kiniksa shall (x) use diligent efforts to enable GNE, at its option, to participate in any discussions with Biogen to attempt to resolve any APA Breach Claims and, to the extent an APA Breach Claim becomes subject to judicial, arbitral, mediation or other dispute resolution processes and as permitted by Applicable Law, if requested by GNE, join GNE, and not object to the intervention by, or joinder of, GNE, as a party-in-interest in any such proceedings related thereto; (y) seek to cure or as appropriate dispute any APA Breach Claims as permitted under Section 8.2.3 of the Biogen APA; and (z) to the extent that GNE would be capable of curing any APA Breach Claim relating to the subject matter of this Agreement, at GNE's sole discretion and request, permit GNE on Kiniksa's behalf (and make all diligent efforts with Biogen for it to accept) to cure such APA Breach Claim under the Biogen APA. GNE shall be permitted to offset its reasonable, documented, undisputed costs (including any financial payments made to Biogen therefor) incurred curing such APA Breach Claim against any payments owed to Kiniksa under this Agreement except to the extent such breach of the Biogen APA is directly caused by the actions or omissions of the GNE Group (or, if such costs are in dispute, then GNE may so offset such costs to the extent determined by a court of competent jurisdiction consistent with Section 20.2 (Disputes) and Section 20.3 (Jurisdiction; Consent to Forum)).

14.3 *Representations and Warranties of GNE*

GNE represents and warrants to Kiniksa as of the Execution Date that the execution, delivery and performance of this Agreement and the Biogen Side Letter by it and all instruments and documents to be delivered by it hereunder: (i) are within its corporate power; (ii) have been duly authorized by all necessary or proper corporate action; (iii) are not in contravention of any provision of its certificate of formation or limited liability company agreement; (iv) to its knowledge of, will not violate any law or regulation or any order or decree of any Governmental Authority; (v) will not violate the terms of any indenture, mortgage, deed of trust, lease, agreement, or other instrument to which it is a party or by which it or any of its property is bound, which violation would have an adverse effect on its financial condition or on its ability to perform its obligations hereunder; and (vi) do not require any filing or registration with, or the consent or approval of, any Governmental Authority or any other person, which has not been made or obtained previously (other than approvals required under Antitrust Laws, Marketing Authorizations required for the sale of Licensed Products and filings with Regulatory Authorities required in connection with Licensed Products). Further, GNE represents and warrants to Kiniksa that there are no claims or investigations, pending or threatened against GNE or any of its Affiliates, at law or in equity, or before or by any Governmental Authority relating to the matters contemplated under this Agreement or the Biogen Side Letter, or that would materially adversely affect GNE's ability to perform its obligations hereunder or thereunder, and neither GNE nor any of its Affiliates is or will be under any obligation to any person, contractual or otherwise, that is conflicting with the terms of this Agreement or the Biogen Side Letter, or that would impede the fulfillment of GNE's obligations hereunder or thereunder.

14.4 *No Other Representations and Warranties*

EXCEPT AS OTHERWISE PROVIDED IN THIS AGREEMENT, THE FOREGOING REPRESENTATIONS AND WARRANTIES ARE IN LIEU OF ALL OTHER REPRESENTATIONS AND WARRANTIES, EXPRESS OR IMPLIED, INCLUDING WARRANTIES OF MERCHANTABILITY OR FITNESS FOR A PARTICULAR PURPOSE OF PRODUCTS.

15. Indemnification

15.1 *Indemnification by GNE*
GNE shall indemnify, hold harmless and defend Kiniksa, Kiniksa's Affiliates and their directors, officers, employees and agents ("**Kiniksa Indemnitees**") from and against any and all losses, expenses, cost of defense (including reasonable attorneys' fees, witness fees, damages, judgments, fines and amounts paid in settlement) incurred in connection with any claims, demands, actions or other proceedings by any Third Party and any amounts Kiniksa Indemnitees become legally obligated to pay to such Third Party, to the extent that such claim or claims arise out of or result from (a) any breach of this Agreement by the GNE Group, (b) the development, manufacture, or commercialization of any Licensed Antibody or Licensed Product (*e.g.*, product liability claims) by or on behalf of the GNE Group, or (c) the negligence or willful misconduct by or on behalf of the GNE Group, or any of its respective directors, officers, employees, subcontractors, or agents in the performance of GNE's obligations or exercise of its rights under this Agreement, except, in each case ((a) through (c)), to the extent such losses, expenses, costs and amounts arise out of or result from the matters described under Section 15.2 (Indemnification by Kiniksa).

15.2 *Indemnification by Kiniksa*
Kiniksa shall indemnify, hold harmless and defend GNE, GNE's Affiliates and their directors, officers, employees and agents ("**GNE Indemnitees**") from and against any and all losses, expenses, cost of defense (including reasonable attorneys' fees, witness fees, damages, judgments, fines and amounts paid in settlement) incurred in connection with any claims, demands, actions or other proceedings by any Third Party and any amounts GNE Indemnitees become legally obligated to pay to such Third Party, to the extent that such claim or claims arise out of or result from (a) any breach of this Agreement by Kiniksa; (b) the development, manufacture, or commercialization of any Licensed Antibody or Licensed Product (*e.g.*, product liability claims) by or on behalf of Kiniksa or any of its Affiliates or sublicensees, or (c) the negligence or willful misconduct by or on behalf of Kiniksa or any of its Affiliates or sublicensees, or any of their respective directors, officers, employees, subcontractors, or agents in the performance of Kiniksa's obligations or exercise of its rights under this Agreement, except in each case ((a) through (c)) to the extent such losses, expenses, costs and amounts arise out of or result from the matters described under Section 15.1 (Indemnification by GNE).

15.3 *Procedure*
In the event of a claim by a Third Party against a Party entitled to indemnification under this Agreement ("**Indemnified Party**"), the Indemnified Party shall promptly notify the other Party ("**Indemnifying Party**") in writing of the claim and the Indemnifying Party shall undertake and solely manage and control, at its sole expense, the defense of the claim and its settlement. The Indemnified Party shall cooperate with the Indemnifying Party and may, at its option and expense, be represented in any such action or proceeding by counsel of its choice. The Indemnifying Party shall not be liable for any litigation costs or expenses incurred by the Indemnified Party without the Indemnifying Party's written consent. The Indemnifying Party shall not settle any such claim unless: (a) such compromise or settlement imposes only a monetary obligation on the Indemnifying Party and includes as an unconditional term thereof the giving by each claimant or plaintiff of the Indemnified Party a release from all liability in respect of such claim; or (b) the Indemnified Party consents to such compromise or settlement, which consent will not be unreasonably withheld unless such compromise or settlement involves (i) any admission of legal wrongdoing by the Indemnified Party, (ii) any payment by the Indemnified Party that is not indemnified under this Agreement, or (iii) the imposition of any equitable relief

against the Indemnified Party (in which case, (i) through (iii), the Indemnified Party may withhold its consent to such settlement in its sole discretion).

16. Liability

16.1 *Limitation of Liability*

EXCEPT FOR INDEMNIFICATION UNDER ARTICLE 15 (INDEMNIFICATION), ANY BREACH OF THE PARTIES' OBLIGATIONS UNDER ARTICLE 17 (CONFIDENTIAL INFORMATION), OR ANY MISAPPROPRIATION, INFRINGEMENT OR OTHER VIOLATION OF INTELLECTUAL PROPERTY OWNED OR CONTROLLED BY THE OTHER PARTY, NEITHER PARTY SHALL BE ENTITLED TO RECOVER FROM THE OTHER PARTY ANY SPECIAL, INCIDENTAL, CONSEQUENTIAL OR PUNITIVE DAMAGES IN CONNECTION WITH THIS AGREEMENT OR ANY LICENSE GRANTED HEREUNDER. NOTWITHSTANDING THE FOREGOING, NOTHING IN THIS SECTION 16.1 (LIMITATION OF LIABILITY) IS INTENDED TO OR SHALL LIMIT OR RESTRICT ANY DAMAGES TO THE EXTENT ARISING FROM OR RELATING TO WILLFUL MISCONDUCT OR FRAUDULENT ACTS OR FRAUDULENT OMISSIONS OF EITHER PARTY.

17. Confidential Information

17.1 *Confidential Information*

The Licensed Know-How will be the Confidential Information of Kiniksa (for clarity, except to the extent that such Licensed Know-How is excluded from being Confidential Information in accordance with Section 1.38 (i) through (v)). Any report or other information provided by GNE to Kiniksa under this Agreement will be the Confidential Information of GNE. The Joint Know-How and the terms of this Agreement shall be considered Confidential Information of the Parties.

17.2 *Non-Use and Non-Disclosure*

Commencing on the Execution Date and during the Agreement Term and for [***] thereafter, subject to the provisions of this Article 17 (Confidential Information), a Receiving Party shall (i) except to the extent authorized by this Agreement, maintain in confidence and otherwise safeguard, and not published or otherwise disclose, all Confidential Information of a Disclosing Party, (ii) treat Confidential Information provided by Disclosing Party as it would treat its own information of a similar nature, (iii) take all reasonable precautions not to disclose such Confidential Information to Third Parties, without the Disclosing Party's prior written consent, and (iv) not use such Confidential Information other than for fulfilling its obligations under this Agreement. Each Party will promptly notify the other Party of any misuse or unauthorized disclosure of the other Party's Confidential Information.

17.3 *Permitted Disclosure*

Notwithstanding the obligation of non-use and non-disclosure set forth in Section 17.2 (Non-Use and Non-Disclosure), the Parties recognize the need for certain exceptions to this obligation, specifically set forth in Section 17.4 (Press Releases), Section 17.5 (Publications), Section 17.6 (Commercial Considerations), and Section 17.7 (Court or Administrative Order; Compliance with Law) below, with respect to press releases, patent rights, publications, certain commercial considerations or court or administrative order, respectively.

17.4 *Press Releases*

Kiniksa may issue a press release announcing the existence and selected key terms of this Agreement, in the form of the approved press release attached as Appendix 17.4.

GNE may issue press releases in accordance with its internal policy and the terms and conditions of this Agreement. If GNE intends to make reference to Kiniksa in any press release (other than naming Kiniksa as a licensor for Licensed Antibody or Licensed Product under this Agreement), or disclose Confidential Information of Kiniksa not otherwise permitted under the terms of this Agreement, GNE shall provide Kiniksa with a copy of any draft press release related to the activities contemplated by this Agreement at least [***] Business Days prior to its intended publication for Kiniksa's review. Kiniksa may provide GNE with suggested modification to the draft press release. GNE shall consider Kiniksa's suggestions in issuing its press release and will, if so suggested by Kiniksa, remove from such release, prior to its disclosure, any Confidential Information of Kiniksa's that Kiniksa identifies therein.

Kiniksa shall only issue press releases related to the activities contemplated by this Agreement that have been approved by GNE in accordance with, and to the extent required by, this Section 17.4 (Press Releases) or as otherwise permitted under Section 17.7 (Court or Administrative Order; Compliance with Law). In all circumstances, to the extent practicable, Kiniksa shall provide GNE with a draft press release at least [***] Business Days prior to its intended publication for GNE's review. During such period, GNE shall (i) approve the draft press release and permit Kiniksa to issue the press release, (ii) contact Kiniksa to discuss modification to the draft press release, or (iii) contact Kiniksa and disapprove the press release. If GNE asks for modification, then Kiniksa shall either make such modification or work with GNE to arrive at a press release that GNE approves. Notwithstanding any provision to the contrary set forth in this Agreement, after issuance of a press release by a Party in accordance with this Section 17.4 (Press Releases), each Party may further disclose the information contained in such press release without the need for further notice to, or review by, the other Party so long as such information remain accurate and current; *provided* that Kiniksa shall use reasonable efforts to notify GNE prior to the issuance of any such press release as described in this sentence.

To ensure communication alignment, responses (if any) to inquiries by media or other Third Parties after issuance of a permitted press release by Kiniksa (solely or jointly with GNE) shall consist solely of the press release language or shall follow the response guidelines that may be developed by agreement of the Parties.

17.5 *Publications*

During the Agreement Term, the following restrictions shall apply with respect to disclosure by any Party of Confidential Information relating to the Licensed Product in any publication or presentation:

a) GNE, in accordance with its internal policies and procedures, shall have the right to publish all studies, clinical trials and results thereof relating to Licensed Antibodies or Licensed Products (including any results relating to the In Progress PN Study when available and any Joint Know-How) at its discretion, including on the clinical trial registries that are maintained by or on behalf of GNE. Kiniksa shall not publish any studies, clinical trials or results thereof related to Licensed Antibodies or Licensed Product on its clinical trial registry or elsewhere; *provided* that, if Kiniksa is required under Applicable Law to post or otherwise disclose results with respect to the In Progress PN Study, then Kiniksa will post or disclose such study, subject to GNE prior review and comment.

b) Subject to clause (a) above, if GNE intends to include any Confidential Information of Kiniksa (that it is not otherwise permitted to disclose under this Agreement) in a publication

or presentation, then GNE shall provide Kiniksa with a copy of any proposed publication or presentation under this Section 17.5 (Publications) at least [***] days prior to submission for publication ([***] days for poster or abstract presentations) to provide Kiniksa with an opportunity to recommend any changes it reasonably believes are necessary to continue to maintain confidentiality of Kiniksa's Confidential Information identified in such publication or presentation in accordance with the requirements of this Agreement. GNE shall implement Kiniksa's reasonable suggestions for purposes of protecting the confidentiality of Kiniksa's Confidential Information in issuing its publication or presentation.

17.6 *Commercial Considerations*

Each Party and its Affiliates may provide or permit access to the other Party's Confidential Information to the Receiving Party's employees, consultants, advisors, licensees, collaboration partners, and Sublicensees, and to the employees, consultants and advisors of the Receiving Party's Affiliates as necessary or appropriate to conduct such Party's activities (or exercise its rights) under this Agreement who are subject to obligations of confidentiality and non-use with respect to such Confidential Information no less stringent than the obligations of confidentiality and non-use of the Receiving Party pursuant to this Article 17 (Confidential Information). Each Party will remain responsible for any failure by its Affiliates, licensees, collaboration partners, or Sublicensees, and its and its Affiliates' respective employees, consultants and advisors, to treat such Confidential Information as required under this Article 17 (Confidential Information) as if such Affiliates, employees, consultants, advisors, licensees, collaboration partners, and Sublicensees were parties directly bound to the requirements of this Article 17 (Confidential Information).

Notwithstanding the obligation of non-use and non-disclosure set forth in Section 17.2 (Non-Use and Non-Disclosure), (a) GNE or its Affiliates may disclose Confidential Information of Kiniksa (including Joint Know-How) to (i) Governmental Authorities to the extent required or desirable to secure government approval for the development, manufacture or sale of Licensed Product in the Territory, (ii) Third Parties acting on behalf of GNE, to the extent reasonably necessary for the development, manufacture, marketing, or sale of Licensed Product in the Territory, *provided* that such disclosure is covered by terms of confidentiality substantially similar as those set forth herein, (iii) Third Parties requesting clinical trial data information (in accordance with GNE's then-current data sharing policy), or (iv) Third Parties to the extent reasonably necessary to market the Licensed Product; and (b) the Receiving Party may disclose Confidential Information of the Disclosing Party to the Receiving Party's directors, advisors (including financial advisors, attorneys and accountants), actual or *bona fide* potential acquisition partners, potential financing sources or investors, and underwriters on a need to know basis; *provided* that (I) such disclosure is covered by terms of confidentiality similar to those set forth herein (which may include professional statutory obligations), (II) such disclosure is limited to the maximum extent practicable for the particular context in which it is being disclosed, and (III) the term of such confidentiality obligation must be consistent with industry standards.

17.7 *Court or Administrative Order; Compliance with Law*

Notwithstanding the obligation of non-use and non-disclosure set forth in Section 17.2 (Non-Use and Non-Disclosure) or the approval requirements related to press releases set forth in Section 17.4 (Press Releases), the Receiving Party or its Affiliates may disclose the Confidential Information of the Disclosing Party as and to the extent that such Confidential Information is required to be disclosed by the Receiving Party or its Affiliates to comply with a court or administrative order, Applicable Law, IFRS, GAAP, applicable regulations of a stock exchange, or to defend or prosecute litigation; *provided* that (a) the Receiving Party or its Affiliates

furnishes prompt notice (in no event less than [***] days, or such shorter period as may be required to comply with regulation of any stock exchange or other Applicable Law, order, or regulation and in no event will the disclosing Party be required to delay any filing or disclosure required under Applicable Law (including SEC regulations)) of such disclosure to the Disclosing Party to enable it to resist such disclosure and, to the extent practicable, takes reasonable and lawful actions to minimize the degree of such disclosure, including seeking a protective order or other appropriate remedy (including redaction) to ensure confidential treatment of such information; and (b) upon GNE's request Kiniksa shall provide an explanation of any legal advice it received that Kiniksa is required to make such disclosures. No notice shall be required under this Section 17.7 (Court or Administrative Order; Compliance with Law) if and to the extent that the specific information contained in the proposed disclosure has previously been included in any previous disclosure made by either Party hereunder pursuant to this Article 17 (Confidential Information), or is otherwise approved in advance in writing by the other Party.

In addition, either or both Parties may be obligated to make a filing or disclosure of a copy of this Agreement (including any subsequent amendments thereto) with the SEC (or equivalent foreign agency) or a Governmental Authority, and each Party shall be entitled to make such a required filing or disclosure; *provided* that, to the extent not prohibited by Applicable Law or judicial or administrative process, prior to making any such filing or disclosure, such Party shall provide a draft of this Agreement (or amendments thereto, as applicable) to the other Party at least [***] Business Days in advance of such filing or disclosure to provide the other Party the opportunity to review and comment and shall specify to the non-disclosing Party when its comments need to be provided in order to be considered. The non-disclosing Party shall provide any comments within [***] Business Days, and the disclosing Party shall consider in good faith any timely comments provided by the non-disclosing Party; *provided* that the disclosing Party may or may not accept such comments in its discretion based upon the advice of its legal counsel as to the legal necessity to reject such comments for such filing (provided that upon GNE's request Kiniksa shall provide an explanation of such legal advice) and in no event will the disclosing Party be required to delay any filing or disclosure required under Applicable Law (including SEC regulations). Each Party shall be responsible for its own legal and other external costs in connection with any such filing or disclosure pursuant to this paragraph.

18. Term and Termination

18.1 *Commencement and Term*
This Agreement shall commence upon the Effective Date and continue for the Agreement Term; *provided* that certain terms shall be effective and commence as of the Execution Date in accordance with Section 20.18.4 (Provisions Effective on Execution Date). Following the expiration of the Agreement Term for a given Licensed Product in a given country, the licenses granted to GNE under Section 2.1 (Licenses) for such Licensed Product in such country shall be fully paid up, irrevocable and royalty-free.

18.2 *Termination*

18.2.1 Termination for Breach
A Party ("**Non-Breaching Party**") shall have the right to terminate this Agreement in its entirety in the event the other Party ("**Breaching Party**") is in material breach of its material obligations under this Agreement. The Non-Breaching Party shall provide written notice to the Breaching Party, which notice shall identify in reasonable detail the nature of the breach. The Breaching

Party shall have a period of [***] after such written notice is provided ("**Peremptory Notice Period**") to cure such breach. If the Breaching Party has a *bona fide* dispute as to whether such breach occurred or has been cured, or whether it is a breach of any of its material obligations under this Agreement, it will so notify the Non-Breaching Party, and the expiration of the Peremptory Notice Period shall be tolled until such dispute is resolved pursuant to Section 20.2 (Disputes) and Section 20.3 (Jurisdiction; Consent to Forum). Upon a determination of breach of any of its material obligations under this Agreement or failure to cure such breach, the Breaching Party will have the remainder of the Peremptory Notice Period, if any, to cure such breach. If such breach is not cured within the Peremptory Notice Period, then absent withdrawal of the Non-Breaching Party's request for termination and, if such termination right is for a material breach by Kiniksa subject to GNE's right under Section 18.5 (GNE Rights in Lieu of Termination For Material Breach by Kiniksa), this Agreement shall terminate in its entirety effective as of the expiration of the Peremptory Notice Period.

Any failure to comply by GNE with GNE's reporting obligations under Section 4.5.1 (Records; Reports) or Section 4.5.2 (Content of Reports), as described in Section 4.5.3 (Inability to Report Specified Details Not a Material Breach), shall not be considered a material breach by GNE for purposes of giving a right to Kiniksa to terminate this Agreement, in whole or in part, under this Section 18.2.1 (Termination for Breach).

18.2.2 Termination for Biogen Reversion

Upon the occurrence of a Biogen Reversion, subject to Section 18.6.5 (License to GNE Upon Biogen Reversion), this Agreement shall terminate effective upon the effective date of the termination of the Biogen APA.

18.2.3 Termination by GNE for Effective Date Rep Failure

Upon the occurrence of a Rep Failure that GNE has not waived in writing in accordance with Section 20.18.3 (Effective Date; Effective Date Representations and Warranties), GNE shall have the right to terminate this Agreement in the entirety by written notice to Kiniksa specifying such a termination under this Section 18.2.3 (Termination by GNE for Effective Date Rep Failure). Such termination shall be effective immediately upon GNE's delivery of such written notice and, for clarity, in such case the Effective Date shall be deemed to not have occurred.

18.3 *Insolvency*

A Party shall have the right to terminate this Agreement in the entirety if the other Party incurs an Insolvency Event; *provided*, *however*, that in the case of any involuntary bankruptcy proceeding, such right to terminate shall only become effective if the Party that incurs the Insolvency Event consents to the involuntary bankruptcy or such proceeding is not dismissed within [***] days after the filing thereof.

18.4 *Termination by GNE without a Cause*

GNE shall have the right to terminate this Agreement at any time in its entirety upon (i) [***] prior written notice, if such notice is delivered prior to the First Commercial Sale of a Licensed Product; or (ii) [***] prior written notice, if such notice is delivered after the First Commercial Sale of a Licensed Product. The effective date of termination under this Section 18.4 (Termination by GNE without a Cause) shall be the date [***] or [***], as the case may be, after GNE provides such written notice to Kiniksa.

18.5 *GNE Rights in Lieu of Termination For Material Breach by Kiniksa*

In the event that GNE has the right to terminate this Agreement pursuant to Section 18.2.1 (Termination for Breach), following the expiration of all applicable notice and cure periods, and, if any dispute is initiated under Sections 20.2 (Disputes) and 20.3 (Jurisdiction; Consent to Forum) before the expiration of the applicable cure period with respect to the asserted basis of such termination, agreement by the Parties or confirmation by a court of competent jurisdiction consistent with Section 20.3 (Jurisdiction; Consent to Forum) of the basis for termination under Section 18.2.1 (Termination for Breach), GNE may elect, at its sole option, upon written notice to Kiniksa that, in lieu of exercising its right to terminate this Agreement pursuant to Section 18.2.1 (Termination for Breach), this Agreement shall remain in full force and effect; *provided*, *however*, that, (a) to the extent not already paid by GNE under this Agreement, GNE shall remain obligated to pay milestones and other payments in accordance with Sections 9.2 (Fee upon Satisfaction of Clinical Supply by Required Supply Dates), 9.3 (Development and Regulatory Event Payments) and 9.4 (Sales Based Events) and royalties under Section 9.5 (Royalty Payments); and (b) GNE shall have the right to offset the full amount of any undisputed damages incurred by GNE as a result of such material breach by Kiniksa from any future payments otherwise due and payable to Kiniksa under this Agreement (or, if such damages are in dispute, then GNE may so offset those damages incurred by GNE as a result of such material breach by Kiniksa to the extent determined by a court of competent jurisdiction consistent with Section 18.2.1 (Termination for Breach)), or as otherwise payable by GNE to Biogen in accordance with the Biogen Side Letter.

18.6 *Consequences of Termination*

18.6.1 Effects of Termination

Upon any termination of this Agreement under (i) Section 18.2 (Termination) (except to the extent specified in Section 18.6.5 (License to GNE Upon Biogen Reversion)), (ii) Section 18.3 (Insolvency), or (iii) Section 18.4 (Termination by GNE without a Cause), the rights and licenses granted by any Party to the other Party under this Agreement shall terminate in their entirety on the effective date of such termination.

Upon any termination of this Agreement in the entirety under Section 18.2.1 (Termination for Breach) or Section 18.4 (Termination by GNE without a Cause), if Kiniksa desires to continue development or commercialization of the Licensed Antibodies and Licensed Product that were developed or commercialized by GNE under this Agreement (the "**Reversion Product(s)**"), then Kiniksa shall provide a Continuation Election Notice to GNE no later than [***] days following, as applicable (a) the effective date of a termination pursuant to Section 18.2.1 (Termination for Breach); or (b) the notice of a termination pursuant to Section 18.4 (Termination by GNE without a Cause). If GNE receives such a Continuation Election Notice:

(a) After the effective date of termination GNE shall, to the extent GNE has the right to do so, transfer to Kiniksa all Regulatory Filings and approvals, all final pre-clinical and clinical study reports and clinical study protocols, and all data that is GNE Know-How, including clinical data, in GNE's Control specifically related to such Reversion Product(s) necessary for Kiniksa to continue to develop, manufacture, and commercialize such Reversion Product(s). All data shall be transferred in the form and format in which it is maintained by GNE. Original paper copies shall only be transferred if legally required or where generating electronic copies is not reasonably feasible. GNE shall not be required to prepare or finalize any new data, reports or information solely for purposes of transfer to Kiniksa.

(b) GNE shall assign all agreements solely related to such Reversion Products, to the extent such agreements have not been cancelled and are assignable without GNE paying any consideration (unless Kiniksa agrees to bear any such consideration) or commencing litigation in order to effect an assignment of any such agreement.

(c) Kiniksa shall, upon transfer, have the right to disclose such filings, approvals and data to (i) Governmental Authorities to the extent required or desirable to secure approval for the development, manufacture or sale of such Reversion Product(s); (ii) Third Parties acting on behalf of Kiniksa, its Affiliates or licensees, to the extent necessary solely for the development, manufacture, or commercialization of such Reversion Product(s); or (iii) Third Parties to the extent reasonably necessary to develop, manufacture, or commercialize such Reversion Product(s).

(d) GNE shall grant to Kiniksa, an exclusive, sub-licensable (through multiple tiers) license under the GNE Patent Rights and GNE Know-How, including GNE's interest in the Joint Patent Rights and Joint Know-How, solely to the extent necessary to allow Kiniksa, its Affiliates or licensees to develop, subject to Section 18.6.3.2(b) (Obligations Related to Manufacturing) and Section 18.6.3.3 (Limitations on Grant-Backs; Transfer Expenses) manufacture and have manufactured, use, offer to sell, sell, promote, export and import such Reversion Product(s). For clarity, the licenses under this Section 18.6.1(d) shall not include any licenses that GNE has with a Third Party for which such grant would be prohibited or under which a member of the GNE Group would incur financial obligations to such Third Party, unless Kiniksa agrees to bear any such consideration. Such license shall be subject to, and under financial terms to be negotiated in good faith between the Parties, taking into account the value of such GNE Patent Rights and GNE Know-How and the contributions made by GNE to the development, commercialization and manufacture of such Reversion Product(s), and the circumstances of termination of the Agreement. If the Parties fail to reach agreement on the financial terms of such license within [***] days following GNE's timely receipt of a Continuation Election Notice, or a later date agreed to by each of the Parties, then such dispute shall be referred to the officers of the Parties for resolution. If the Parties' officers do not fully resolve such matter within [***] Business Days (or a later date agreed to by each of the Parties) (the **"Reversion Negotiation Expiration"**), then such financial terms shall be subject to final resolution by baseball binding arbitration pursuant to the terms set forth on Appendix 18.6.1(d).

18.6.2 Direct License

Notwithstanding any provision to the contrary in this Agreement, upon a termination of this Agreement in the entirety (except for a termination of this Agreement under Section 18.2.2 (Termination for Biogen Reversion) (if GNE receives the Stand-By License (as defined in the Biogen Side Letter) from Biogen under the Biogen Side Letter) or Section 18.2.3 (Termination by GNE for Effective Date Rep Failure)): (a) any Compulsory Sublicense shall remain in full force and effect as may be required by Applicable Law and, to the extent permitted by Applicable Law, GNE shall assign such Compulsory Sublicense to Kiniksa; and (b) upon the written request of any permitted Sublicensee under a valid sublicense agreement granted by GNE pursuant to Section 2.2 (Sublicense), Kiniksa agrees to enter into a direct license grant to such Sublicensee consistent in scope with the rights sublicensed to such Sublicensee under this Agreement; *provided* that (i) such permitted Sublicensee is not then in breach of its sublicense agreement and did not cause any breach by GNE that gave rise to a termination of this Agreement by Kiniksa under Section 18.2.1 (Termination for Breach) and (ii) agrees to be bound by all applicable terms and conditions of this Agreement, including rendering directly to

Kiniksa all payments and other obligations due to Kiniksa related to such sublicense agreement (including all event payments and royalty payments).

18.6.3 Other Obligations

Upon a termination of this Agreement in the entirety (except for a termination of this Agreement under Section 18.2.2 (Termination for Biogen Reversion) (if GNE receives the Stand-By License (as defined in the Biogen Side Letter) from Biogen under the Biogen Side Letter) or Section 18.2.3 (Termination by GNE for Effective Date Rep Failure)):

18.6.3.1 Obligations Related to Ongoing Activities

If Kiniksa does not provide timely Continuation Election Notice, then GNE (a) shall have the right to cancel all ongoing obligations and (b) shall complete all non-cancellable obligations at its own expense.

If Kiniksa provides such timely Continuation Election Notice, then from the date of notice of termination until the effective date of termination, with respect to the Reversion Products, GNE shall continue activities, including preparatory activities, ongoing as of the date of notice of termination. However, GNE shall not be obliged to initiate any new activities not ongoing at the date of notice of termination.

After the effective date of termination, GNE shall have no obligation to perform or complete any activities or to make any payments for performing or completing any activities under this Agreement, except as expressly stated herein.

Notwithstanding the foregoing, upon the request of Kiniksa, GNE shall complete any Clinical Studies related to the Reversion Product(s) that are being conducted under its IND for such Reversion Product(s) and are ongoing as of the effective date of termination; *provided*, *however*, that (i) both Kiniksa and GNE in their reasonable judgment have concluded that completing any such Clinical Studies does not present an unreasonable risk to patient safety; (ii) GNE shall have no obligation to recruit or enroll any additional patients after the effective date of termination; and (iii) Kiniksa agrees to reimburse GNE for all of its development costs that arise after the effective date of termination in completing such Clinical Studies.

18.6.3.2 Obligations Related to Manufacturing

(a) Clinical Supplies
Upon Kiniksa's request, GNE shall transfer all existing and available clinical material solely relating to Reversion Products to Kiniksa at [***]. GNE shall have no obligation to perform any additional activities concerning the clinical supplies (*e.g.*, retesting, analyses). Upon such transfer Kiniksa shall assume all liability for the use of such material.

(b) Commercial Supplies
Upon the request of Kiniksa, with respect to the Reversion Product, GNE shall (itself or through its Third Party contractor), if necessary during the transition of manufacturing responsibilities back to Kiniksa or its designee, manufacture and supply reasonable amounts of such Reversion Product to Kiniksa under a manufacturing transfer and transition plan for a period that shall not exceed [***] from the effective date of the termination of this Agreement at a price to be agreed by the Parties in good faith, but in no event exceeding [***], whether such Reversion Product is manufactured for GNE through a Third Party contract or by GNE itself (or through its Affiliates), as calculated on a consistent basis

according to its then current accounting procedures. Kiniksa shall use Commercially Reasonable Efforts to take over the manufacturing as soon as possible after the effective date of termination.

Subject to Section 18.6.3.3(c) (Limitations on Grant-Backs; Transfer Expenses), if GNE is unwilling to transfer proprietary cell lines, growth media, culture media or disclose proprietary technical development/manufacturing know-how that is necessary to continue manufacturing of a commercialized Reversion Product to the extent applicable to such termination, then GNE may elect to either supply Kiniksa (i) indefinitely on terms to be negotiated, or (ii) until such time as GNE successfully transfers the process to a Third Party contract manufacturing organization acceptable to GNE.

18.6.3.3 Limitations on Grant-Backs; Transfer Expenses

(a) Irrespective of any provision to the contrary in Section 18.6 (Consequences of Termination), all transfers and licenses from GNE to Kiniksa (or other obligations of GNE) under Section 18.6 (Consequences of Termination) are solely with respect to Reversion Product(s) to the extent applicable to such termination that are not Combination Product(s) or Diagnostic Product(s). Such transfers, licenses and obligations do not extend to other therapeutically active ingredients or products, even if physically mixed, combined or packaged together with a Reversion Product, and even if a Reversion Product is intended (according to the investigation plan, proposed labeling or actual labeling, as applicable) for use with such other therapeutically active ingredients or products.

(b) Irrespective of any provision to the contrary in Section 18.6 (Consequences of Termination), in connection with research studies, clinical trials or other activities associated with the development and commercialization of Reversion Products to the extent applicable to such termination, GNE may have collected (i) personally identifiable information about individual human subjects or (ii) human biological samples (collectively, "**PII/Samples**"). Legal and contractual restrictions may apply to such PII/Samples. GNE shall have no obligation to transfer such PII/Samples unless necessary for the continued development of the Reversion Product, in which case GNE shall not be obliged to transfer any PII/Samples that GNE in good faith believes, based on the advice of legal counsel, would be prohibited or would subject GNE to potential liability by reason of Applicable Law, contractual restrictions or insufficient patient consent. If GNE transfers any such PII/Samples, the Parties will enter into the relevant agreements under applicable data privacy laws (such as a data transfer agreement) when required in accordance with Section 7.3 (Data Privacy). Upon the transfer of such PII/Samples by GNE, Kiniksa shall use such PII/Samples for the sole purpose of developing and commercializing the Reversion Product, and Kiniksa shall be responsible for the correct and lawful use of the PII/Sample in compliance with the applicable data protection laws, the informed consent forms and privacy notices (including but not limited to potential re-consenting of the patients at Kiniksa's costs if the legal basis for the processing of the patients' data was their explicit consent).

(3) Except as expressly required under Section 18.6.3.2(b), with respect to Reversion Products that are biologics, GNE shall be under no obligation to provide proprietary cell lines, growth media, culture media, or disclose proprietary technical development/manufacturing know-how, except, upon Kiniksa's request, the Parties will discuss in good faith to what extent and under which conditions (which at GNE's option may include only providing to a CMO approved by GNE) GNE will provide to Kiniksa such proprietary cell lines, growth media, culture media, or disclose proprietary technical development/manufacturing know-how that is necessary for Kiniksa continue the development or manufacture of Reversion Products.

(4) Irrespective of any provision to the contrary in Section 18.6 (Consequences of Termination), Kiniksa shall promptly reimburse GNE for [***] incurred by or on behalf of GNE for transfer activities from GNE to Kiniksa under Section 18.6.1 (Effects of Termination) ("**GNE Transfer Activities**"), except that transfer activities corresponding to the return of material remains, data, reports, records, documents, Regulatory Filings and Marketing Authorizations originally provided by Kiniksa to GNE ("**Kiniksa-Originated Transfer Activities**") shall be [***]. If Kiniksa desires GNE Transfer Activities other than Kiniksa-Originated Transfer Activities, then Kiniksa shall make a payment to GNE of [***] US dollars (US$[***]) ("**Minimum Transfer Payment**"); *provided* that such Minimum Transfer Payment need not be paid if this Agreement was terminated by Kiniksa for GNE's material breach under Section 18.2.1 (Termination for Breach) or by GNE under Section 18.4 (Termination by GNE without a cause). The Minimum Transfer Payment shall be non-refundable, but shall be fully creditable against Kiniksa's reimbursement for the GNE Transfer Activities. GNE shall be under no obligation to provide GNE Transfer Activities (beyond than Kiniksa-Originated Transfer Activities) prior to receipt, if applicable pursuant to the foregoing, of the Minimum Transfer Payment or if the Minimum Transfer Payment is received after the effective date of the termination.

(5) Unless otherwise agreed to by the Parties, the transfer of physical materials contemplated under this Section 18.6.3 (Other Obligations), except in conjunction with such future toll manufacturing obligations as may apply under Section 18.6.3.2(b) (Obligations Related to Manufacturing), if applicable, shall be delivered, at GNE's option, [***] (Incoterms 2020).

(6) Notwithstanding any provision to the contrary in this Article 18 (Term and Termination) or elsewhere in this Agreement, no licenses or rights are granted by GNE to Kiniksa under any information, data, proprietary materials or other intellectual property rights whether or not patentable that are owned or controlled by Flatiron Health Inc., a Delaware corporation or Foundation Medicine, Inc., a Delaware corporation.

18.6.4 Royalty and Payment Obligations

Termination of this Agreement by a Party, for any reason, shall not release GNE from any obligation to pay royalties or make any payments to Kiniksa that are accrued prior to the effective date of termination. Termination of this Agreement by a Party, for any reason, will release GNE from any obligation to pay royalties or make any payments to Kiniksa hereunder that would otherwise become payable on or after the effective date of termination.

18.6.5 License to GNE Upon Biogen Reversion

In the event this Agreement is terminated pursuant to Section 18.2.2 (Termination for Biogen Reversion) and such termination is not directly attributable to any breach of this Agreement by any member of the GNE Group, then:

(a) if any Licensed IP remains in Kiniksa's Control after the effective date of termination of the Biogen APA, but prior to Kiniksa taking such actions and executing such documents to assign to Biogen all of its rights, title, and interests in and to the Purchased Assets (as such term is defined in the Biogen APA) pursuant to Section 8.3(c) of the Biogen APA, then until such time as ownership of such Purchased Assets vests in Biogen, Kiniksa hereby grants GNE an exclusive, sublicensable (through multiple tiers), fully paid up, irrevocable and royalty-free license, under any interest Kiniksa may have in the Purchased Assets, to research, have researched, develop, have developed, register, have registered, use, have used, make, have made, import, have imported, export, have exported, market, have marketed, distribute, have distributed, sell and have sold Licensed Antibodies and Licensed

Products in the Field in the Territory; and

(b) if any Licensed IP other than the Purchased Assets (as such term is defined in the Biogen APA) remains in Kiniksa's Control after the effective date of the termination of the Biogen APA, Kiniksa hereby grants GNE a non-exclusive, sublicensable (through multiple tiers), fully paid up, irrevocable and royalty-free license, under any interest Kiniksa may have in such Licensed IP, to research, have researched, develop, have developed, register, have registered, use, have used, make, have made, import, have imported, export, have exported, market, have marketed, distribute, have distributed, sell and have sold Licensed Antibodies and Licensed Products in the Field in the Territory.

18.7 *Survival*
Article 1 (Definitions) (to the extent necessary to interpret this Agreement), Articles 9 (Payment), 10 (Accounting and Reporting) and 12 (Auditing) each to the extent payment obligations exist at the time of termination, Article 11 (Taxes) to the extent such were incurred at the time of termination, Section 13.1 (Ownership of Inventions); Article 15 (Indemnification), Article 16 (Liability), Article 17 (Confidential Information), Article 18 (Term and Termination), and Section 20.1 (Governing Law; Jurisdiction); Section 20.3 (Jurisdiction; Consent to Forum) shall survive any expiration or termination of this Agreement for any reason.

19. Bankruptcy

All licenses (and to the extent applicable rights) granted under or pursuant to this Agreement by Kiniksa to GNE are, and shall otherwise be deemed to be, for purposes of Section 365(n) of Title 11, US Code (the "**Bankruptcy Code**") licenses of rights to "intellectual property" as defined under Section 101(35A) of the Bankruptcy Code. Unless GNE elects to terminate this Agreement, the Parties agree that GNE, as a licensee or sublicensee of such rights under this Agreement, shall retain and may fully exercise all of its rights and elections under the Bankruptcy Code, subject to the continued performance of its obligations under this Agreement.

20. Miscellaneous

20.1 *Governing Law; Jurisdiction.*
This Agreement shall be governed by and construed in accordance with the laws of New York, without reference to its conflict of laws principles. This Agreement and its effect are subject to and shall be construed and enforced in accordance with the law of the State of New York, without regard to its conflicts of laws that would require the application of any other law.

20.2 *Disputes*
In the event of the occurrence of dispute, the Parties shall first refer such dispute to their respective Alliance Managers for attempted resolution by such Alliance Managers within [***] days after such referral. If such dispute is not resolved within such [***] day period, either Kiniksa and Genentech may, by written notice to the other, have such dispute referred to respective officers of the Parties designated below or their designees, for good faith negotiations attempting to resolve the dispute within [***] days after such dispute is referred to such officers, or such longer period as the Parties may agree. The designated executive officers are as follows:

For Kiniksa: CEO
For GNE: Head of Pharma Partnering

20.3 *Jurisdiction; Consent to Forum*

For any dispute under this Agreement that cannot be resolved in accordance with Section 20.2 (Disputes), each of the Parties hereby irrevocably and unconditionally consents to submit to the exclusive jurisdiction of the federal courts located in the Eastern District of the State of New York for any matter arising out of or relating to this Agreement and the transactions contemplated hereby, and agrees not to commence any litigation relating thereto except in such courts. Each of the Parties hereby irrevocably and unconditionally waives any objection to the laying of venue of any matter arising out of this Agreement or the transactions contemplated hereby in the courts of the State of New York and hereby further irrevocably and unconditionally waives and agrees not to plead or claim in any such court that any such matter brought in any such court has been brought in an inconvenient forum. The Parties agree that a final judgment in any such matter shall be conclusive and may be enforced in other jurisdictions by suits on the judgment or in any other manner provided by law. Any proceeding brought by either Party under this Agreement shall be exclusively conducted in the English language.

20.4 *Assignment*

Except as provided in this Section 20.4 (Assignment), neither Party may assign or otherwise transfer this Agreement, or any part thereof (including any rights or obligations hereunder) without the prior written approval of the other Party. Notwithstanding the foregoing, a Party may assign this Agreement in its entirety without such consent (but with prior prompt written notice thereafter), (a) to any of its Affiliates; (b) in the context of a Change of Control of the Party seeking to assign, in which case such Party in its sole discretion may assign its rights and obligations under this Agreement to, as applicable, such Affiliate or acquirer; or (c) in connection with a sale of all or substantially all of the assets to which this Agreement relates; *provided* in each case ((a) - (c)), (i) the person to which this Agreement is so assigned expressly agrees in writing to assume and be bound by all obligations of the assigning Party under this Agreement, and (ii) if Kiniksa is the assigning Party, such person assumes the Biogen APA and is assigned all Licensed IP (and Kiniksa's interest in Joint Patent Rights or the Joint Know-How) and otherwise fully enabled with all the rights of Kiniksa or its Affiliates to perform the obligations of Kiniksa under this Agreement and the Biogen APA. Any permitted assignment shall be binding on the successors of the assigning Party. Any purported assignment in violation of this Section 20.4 (Assignment) will be void and of no force and effect.

20.5 *Debarment*

Kiniksa represents and warrants that neither Kiniksa (or its Affiliates) nor Kiniksa's (or its Affiliates') employees have ever been debarred under 21 U.S.C. §335a, disqualified under 21 C.F.R. §312.70 or §812.119, sanctioned by a Federal Health Care Program (as defined in 42 U.S.C §1320 a-7b(f)), including the federal Medicare or a state Medicaid program, or debarred, suspended, excluded or otherwise declared ineligible from any other similar Federal or state agency or program. In the event Kiniksa (or its Affiliates) or an employee of Kiniksa (or of its Affiliates) that is conducting any Kiniksa Transition Activities, or GNE (or its Affiliates) or an employee of GNE (or of its Affiliates) that is conducting any activities under this Agreement, receives notice of debarment, suspension, sanction, exclusion, ineligibility or disqualification under the above-referenced statutes, then such Party shall ensure the concerned entity or individual shall not continue to perform activities in connection with Licensed Antibodies or Licensed Products, and, in the case of Kiniksa, Kiniksa shall immediately notify GNE in writing of any such debarment.

20.6 *Use of Affiliates*
Each Party may exercise its rights or perform its obligations under this Agreement personally or through one or more of its Affiliates; *provided* that such Party shall nonetheless be primarily liable for the performance of its Affiliates and for any failure by its Affiliates to comply with the restrictions, limitations and obligations set forth in this Agreement and applicable to such Party.

20.7 *Independent Contractor*
No employee or representative of either Party shall have any authority to bind or obligate the other Party to this Agreement for any sum or in any manner whatsoever or to create or impose any contractual or other liability on the other Party without said Party's prior written approval. For all purposes, and notwithstanding any other provision of this Agreement to the contrary, Kiniksa's legal relationship to GNE under this Agreement shall be that of independent contractor, and nothing contained in this Agreement shall be deemed or construed to create a partnership, joint venture, employment, franchise, agency or fiduciary relationship between the Parties.

20.8 *Unenforceable Provisions and Severability*
If any of the provisions of this Agreement are held to be void or unenforceable, then such void or unenforceable provisions shall be replaced by valid and enforceable provisions that will achieve as far as possible the economic business intentions of the Parties. However, the remainder of this Agreement will remain in full force and effect; *provided* that the material interests of the Parties are not affected, *i.e.*, the Parties would presumably have concluded this Agreement without the unenforceable provisions.

20.9 *Waiver*
The failure by either Party to require strict performance or observance of any obligation, term, provision, or condition under this Agreement will neither constitute a waiver thereof nor affect in any way the right of the respective Party to require such performance or observance. The waiver by either Party of a breach of any obligation, term, provision or condition hereunder shall not constitute a waiver of any subsequent breach thereof or of any other obligation, term, provision or condition.

20.10 *Remedies*
No remedy referred to in this Agreement is intended to be exclusive, but each shall be cumulative and in addition to any other remedy referred to in this Agreement or otherwise available under Applicable Law.

20.11 *Interpretation*
Except where the context expressly requires otherwise:

(a) the use of any gender herein shall be deemed to encompass references to either or both genders, and the use of the singular shall be deemed to include the plural (and vice versa),

(b) the words "include", "includes" and "including" shall be deemed to be followed by the phrase "without limitation",

(c) the word "will" shall be construed to have the same meaning and effect as the word "shall",

(d) any definition of or reference to any agreement, instrument or other document herein shall be construed as referring to such agreement, instrument or other document as from time to time amended, supplemented or otherwise modified (subject to any restrictions on such amendments, supplements or modifications set forth herein),

(e) any reference herein to any Party or Third Party or person shall be construed to include the Party's or Third Party's or person's permitted successors and assigns,

(f) the words "herein", "hereof" and "hereunder", and words of similar import, shall be construed to refer to this Agreement in its entirety and not to any particular provision hereof,

(g) all references herein to Articles, Sections, Appendices or Exhibits (as applicable) shall be construed to refer to Articles, Sections, Appendices or Exhibits of this Agreement, and references to this Agreement include all Appendices hereto,

(h) references to any specific law, rule or regulation, or article, section or other division thereof, shall be deemed to include the then-current amendments thereto or any replacement or successor law, rule or regulation thereof, and

(i) the term "or" shall be interpreted in the inclusive sense commonly associated with the term "and/or".

20.12 *Force Majeure*

Neither Party shall be held liable to the other Party nor be deemed to have defaulted under or breached this Agreement for failure or delay in achieving any objective, satisfying any condition, or performing any obligation under this Agreement to the extent, and for so long as, such failure or delay is caused by or results from one or more Force Majeure Events; *provided* that the affected Party gives the other Party prompt written notice of any such Force Majeure Event and the cessation thereof; and *provided further* that the affected Party will notify the other Party in writing of any Force Majeure Event that may affect its performance under this Agreement as soon as reasonably practical, will provide a good faith estimate of the period for which its failure or delay in performance under this Agreement is expected to continue based on currently available information, and will promptly undertake and continues to use reasonable efforts to cure such failure or delay resulting from the Force Majeure Event as soon as practicable and to mitigate its effects, and promptly resumes performance whenever such Force Majeure Event is removed. Any deadline or time period affected by such a Force Majeure Event or a Party's failure to perform resulting therefrom shall be extended automatically by the number of days equal to the number of days that such Force Majeure Event or failure persisted. If a Force Majeure Event persists for more than [***] days, then the Parties will negotiate in good faith any modifications of the terms of this Agreement that may be necessary to arrive at an equitable solution, unless the Party giving such notice has set out a reasonable timeframe and plan to resolve the effects of such force majeure and executes such plan within such timeframe. Notwithstanding the foregoing, the Parties shall not be obliged to negotiate any modification of the terms of this Agreement due to COVID-19 within the first [***] after the Effective Date. Notwithstanding the foregoing, a Party will not be excused from making payments owed hereunder due to any such Force Majeure Event circumstances affecting such Party.

20.13 *Entire Understanding*

This Agreement together with the Biogen Side Letter contains the entire understanding between the Parties hereto with respect to the within subject matter and supersedes any and all prior agreements, understandings and arrangements, whether written or oral including, effective as of the Effective Date, including that certain Non-Disclosure Agreement by and between Hoffmann-La Roche Inc. (an Affiliate of GNE) and Kiniksa effective July 8, 2019, as amended by that certain First Amendment to Non-Disclosure Agreement effective as of May 25, 2021 and that certain Second Amendment to Non-Disclosure Agreement effective as of June 30, 2022 (*provided* that all information disclosed or exchanged under such agreement will be treated as Confidential Information hereunder); *provided* that, if this Agreement is terminated prior to the

Effective Date, then the foregoing referenced Non-Disclosure Agreement shall remain in effect in accordance with its terms.

20.14 *Amendments*
No amendments of the terms and conditions of this Agreement shall be binding upon either Party hereto unless in writing and signed by both Parties.

20.15 *Invoices*
All invoices that are required or permitted hereunder shall be in writing and sent by Kiniksa to GNE at the following address or such other address as GNE may later provide:

[***]

20.16 *Notice*
All notices that are required or permitted hereunder shall be in writing and sufficient if delivered personally, sent by facsimile (and promptly confirmed by personal delivery, registered or certified mail or overnight courier), sent by nationally recognized overnight courier or sent by registered or certified mail, postage prepaid, return receipt requested, addressed as follows:

if to Kiniksa, to:	Kiniksa Pharmaceuticals (UK), Ltd. Third Floor 23 Old Bond Street London, UK, W1S 4PZ Attention: Chief Commercial Officer
with a copy to:	Kiniksa Pharmaceuticals Corp. 100 Hayden Ave. Lexington, MA 02421 United States Attention: General Counsel
and a copy to (which will not constitute notice):	Ropes & Gray LLP 800 Boylston Street; Prudential Tower Boston, MA 02199 Attention: Hannah H. England Email: Hannah.England@ropesgray.com
if to GNE, to:	F. Hoffmann-La Roche Ltd [**]
and:	Genentech, Inc. [**]

or to such other address as the Party to whom notice is to be given may have furnished to the other Party in writing in accordance herewith.

20.17 *Business Day Requirements*

Except as may be otherwise specified in a Pharmacovigilance Agreement or in Section 20.18 (Antitrust and Competition), if any notice or other action or omission is required to be taken by a Party under this Agreement on, or by a day that is not a Business Day, then such notice or other action or omission shall be deemed to be required to be taken on the next occurring Business Day for such Party.

20.18 *Antitrust and Competition*

20.18.1 Antitrust Filing

The Parties shall each, as promptly as practicable after the Execution Date, file or cause to be filed with the FTC, DOJ, and any other applicable Governmental Authority, any Antitrust Filings required under Antitrust Law with respect to the transactions contemplated hereby; *provided* that the Parties shall each make the Antitrust Filing within [***] Business Days after the Execution Date and shall use reasonable efforts to make such Antitrust Filing within [***] Business Days after the Execution Date. The Parties shall use their reasonable best efforts to respond promptly to any requests for additional information made by such agencies, and to cause the waiting period (and any extension thereof) under Antitrust Laws to terminate or expire and resolve any objections by any Governmental Authority at the earliest possible date. For the purposes of this Section 20.18.1 (Antitrust Filing) only, "reasonable best efforts" shall not require either Party to (i) divest, license, sell or hold separate any assets, businesses or properties or enter into any consent decree or assume any other obligations with respect to the ongoing operations of such Party or its Affiliates, or (ii) defend any judicial or administrative litigation. [***].

20.18.2 Cooperation and Assistance

In connection with obtaining clearance under the HSR Act, each Party shall (i) cooperate with each other in connection with any investigation or other inquiry relating to an Antitrust Filing and the transactions contemplated hereby; (ii) keep the other Party or its counsel informed of any communication received from or given to any Governmental Authority relating to the Antitrust Filing and the transactions contemplated hereby (and provide a copy to the other Party if such communication is in writing); (iii) reasonably consult with each other in advance of any meeting or conference with any Governmental Authority, and, to the extent permitted by any Governmental Authority, give the other Party or its counsel the opportunity to attend and participate in such meetings and conferences; and (iv) permit the other Party or its counsel to review in advance, and in good faith consider the views of the other Party or its counsel and incorporating where appropriate, concerning, any submission, filing or communication (and documents submitted therewith) intended to be given to any Governmental Authority.

20.18.3 Effective Date; Effective Date Representations and Warranties

If an Antitrust Filing is made, then, subject to Section 18.2.3 (Termination by GNE for Effective Date Rep Failure), the Effective Date shall occur one Business Day immediately following the Antitrust Clearance Date. Upon the Effective Date, Kiniksa shall deliver to GNE a written certification from an officer of Kiniksa certifying that all the Fundamental Representations remain true and correct as of such date as though then made; *provided* that if (i) Kiniksa is unable or fails to make such a certification as to any Fundamental Representation, or (ii) any Fundamental Representation is no longer true and correct as of such date as though then made on the Execution Date (whether or not notified to GNE as required in this Section 20.18.3 (Effective

Date; Effective Date Representations and Warranties)) (in each case (i) - (ii) a "**Rep Failure**"), then GNE may, in its sole discretion, either (x) in writing waive any such Rep Failure for the purposes of determining the Effective Date, in which case the Effective Date shall occur as otherwise determined by Section 1.49 (Effective Date) or (y) terminate this Agreement pursuant to Section 18.2.3 (Termination by GNE for a Rep Failure Prior to the Effective Date).

Concurrently with the Effective Date (or prior to as the case may be), the Biogen APA Amendment, and each of a certain Asset Transfer Agreement and a certain Side Letter (in each case between Kiniksa and Kiniksa Parent), will become effective in accordance with their terms.

20.18.4 Provisions Effective on Execution Date

This Agreement shall not become effective until the Effective Date (as determined in accordance with Section 20.18.4 (Effective Date; Effective Date Representation and Warranties) and Section 1.49 (Effective Date)) except for the following provisions, which shall become effective as of the Execution Date: Article 1 (Definitions), Article 14 (Representations, Warranties and Covenants), Article 17 (Confidential Information); Section 18.2.3 (Termination by GNE for Effective Date Rep Failure), Section 18.6.1 (Effects of Termination) (solely with respect to the first sentence thereof), and Article 20 (Miscellaneous).

Concurrently with execution and delivery of this Agreement by each of the Parties, the Biogen Side Letter, the Biogen APA Amendment, and each of a certain Asset Transfer Agreement and a certain Side Letter (in each case between Kiniksa and Kiniksa Parent), are each to be executed and delivered by all parties thereto, and, notwithstanding any provision to the contrary in this Agreement, the Execution Date shall not occur unless each such agreement is so executed and delivered by the parties thereto.

20.18.5 Outside Date

This Agreement will terminate (a) at the election of either Party, immediately upon written notice to the other Party, if the FTC or DOJ, or an equivalent authority in the European Union (including, for the avoidance of doubt, the United Kingdom despite it potentially leaving the European Union), seeks a permanent injunction under applicable antitrust and non-competition laws against Kiniksa and GNE to enjoin the transactions contemplated by this Agreement; or (b) at the election of either Party, immediately upon written notice to the other Party, in the event that the Antitrust Clearance Date will not have occurred on or prior to [***] days after the Execution Date, *provided* that the Parties have not agreed in writing to extend the Antitrust Clearance Date. In the event of such termination, all rights and licenses granted hereunder will terminate and this Agreement will be of no further force and effect.

20.19 *Counterparts; e-Signatures*

This Agreement may be executed in any number of counterparts, each of which will be deemed an original, but all of which together will constitute one and the same instrument. For purposes hereof, an e-Signature or email with attached pdf copy, of this Agreement, including the signature pages hereto, will be deemed to be an original. The Parties agree that execution of this Agreement by e-Signatures (as defined below) shall have the same legal force and effect as the exchange of original signatures. Pursuant to this Agreement, "e-Signature" shall mean a signature that consists of one or more letters, characters, numbers or other symbols in digital form incorporated in, attached to or associated with the electronic document, that (a) is unique to the person executing the signature; (b) the technology or process used to make the signature is under the sole control of the person making the signature; (c) the technology or process can be used to identify the person using the technology or process; and (d) the electronic signature

can be linked with an electronic document in such a way that it can be used to determine whether the electronic document has been changed since the electronic signature was incorporated in, attached to or associated with the electronic document.

[Signature Page Follows]

IN WITNESS WHEREOF, the Parties have entered into this Agreement as of the Execution Date.

Kiniksa Pharmaceuticals (UK), Ltd.

/s/ Ross Moat

Name: Ross Moat

Title: Director

F. Hoffmann-La Roche Ltd

/s/ James Sabry

Name: James Sabry

Title: EVP, Global Head, Pharma Partnering

/s/ Peter Trybus

Name: Peter Trybus

Title: Authorized Signatory

Genentech, Inc.

/s/ Edward Harrington

Name: Edward Harrington

Title: CFO Genentech

Appendix 1.96

Kiniksa Development Plan

[***]

Appendix 1.148

Tech Transfer Plan

[***]

Appendix 6.1.2.2

Kiniksa Clinical Supply Obligation for GNE Development

[***]

Appendix 14.1.2

Disclosure Schedule for Section 14.1.2

[***]

Appendix 14.1.4

Licensed IP

[***]

Appendix 17.4

Form of Kiniksa Press Release

Kiniksa Pharmaceuticals Announces Global License Agreement with Genentech for Vixarelimab

– Kiniksa to receive $100 million in upfront and near-term payments –

– Kiniksa is eligible to receive development and commercial milestones as well as royalties on net sales –

– Global license includes development and commercialization rights to vixarelimab –

HAMILTON, BERMUDA – August [X], 2022 – Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) (Kiniksa), a biopharmaceutical company with a portfolio of assets designed to modulate immunological pathways across a spectrum of diseases, today announced a global license agreement with Roche and Genentech, a member of the Roche Group (Genentech), for the rights to develop and commercialize vixarelimab, a fully human monoclonal antibody targeting oncostatin M receptor beta (OSMRβ).

"We are proud to have advanced vixarelimab from a preclinical-stage asset through Phase 2 clinical studies. Our work underscores the differentiated potential of the OSMRβ mechanism as well as its potential to help patients with serious unmet need," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "The agreement provides an optimal infrastructure for the further development of vixarelimab. We plan to allocate the non-dilutive capital received from this transaction towards synergistic opportunities across our portfolio, including the expansion of our ARCALYST cardiovascular franchise."

Under the terms of the global license agreement, Kiniksa will receive $100 million in upfront and near-term payments, and is eligible to receive up to approximately $600 million in certain clinical, regulatory, and sales-based milestones, before fulfilling upstream financial obligations. Kiniksa is also eligible to receive royalties on annual net sales. Genentech will obtain rights for the development and commercialization of vixarelimab. The transaction is subject to certain closing conditions,

including the expiration of the waiting period under the Hart-Scott-Rodino (HSR) Antitrust Improvements Act of 1976 and other customary closing conditions.

Genentech will focus development of vixarelimab in fibrosis, where oncostatin M (OSM)-mediated pathogenesis is thought to be an important pathway for intervention in multiple fibrotic indications.

“Pursuing novel therapies in fibrosis is central to Genentech’s focus on developing medicines for patients with respiratory diseases,” said James Sabry, Global Head of Roche Pharma Partnering. “Developing vixarelimab, a first-in-class fully human monoclonal antibody, in fibrosis is another example of how we are taking an innovative approach to meet patients’ unmet needs.”

Kiniksa has completed screening patients for the Phase 2b clinical trial of vixarelimab in prurigo nodularis. The company plans to complete the trial but will no longer disclose data in the second half of 2022.

About Kiniksa
Kiniksa is a biopharmaceutical company focused on discovering, acquiring, developing, and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa’s portfolio assets, ARCALYST, KPL-404, and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target underserved conditions, and offer the potential for differentiation. These assets are designed to modulate immunological pathways across a spectrum of diseases. For more information, please visit www.kiniksa.com.

About Vixarelimab
Vixarelimab is an investigational fully human monoclonal antibody that targets oncostatin M receptor beta (OSMRβ), which mediates signaling of interleukin-31 (IL-31) and oncostatin M (OSM), two key cytokines implicated in pruritus, inflammation, and fibrosis.

Kiniksa Forward Looking Statements
This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. 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. All statements contained in this press release that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation, statements regarding the licensing of vixarelimab from Kiniksa to Genentech, including (i) anticipated upfront, near-term, milestone and royalty payments under such agreement, (ii) statements regarding the agreement providing an optimal infrastructure for the further development of vixarelimab and (iii) Kiniksa’s plan to allocate the non-dilutive capital received from the transaction towards synergistic opportunities across its portfolio, including the expansion of its ARCALYST cardio-inflammatory franchise; Kiniksa’s plan to complete its Phase 2b clinical trial of

vixarelimab in prurigo nodularis; Genentech's plans for the future development of vixarelimab, including in fibrosis, where oncostatin M (OSM)-mediated pathogenesis is thought to be an important pathway for intervention in multiple fibrotic indications; and Kiniksa's beliefs about the mechanisms of action of vixarelimab and potential impact of its approach in pruritis, inflammation and fibrosis.

These forward-looking statements are based on management's current expectations. These statements are neither promises nor guarantees, but involve known and unknown risks, uncertainties and other important factors that may cause actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements, including without limitation, the following: ours and Genentech's ability to obtain antitrust clearance and close the proposed transaction in a timely manner; Genentech's ability to demonstrate safety and efficacy of vixarelimab in their chosen indications to the satisfaction of applicable regulatory authorities; our ability to realize anticipated near-term payments and milestone and royalty payments under the agreement; our ability to successfully execute on potential synergistic opportunities, including an expansion of our ARCALYST cardio-inflammatory franchise; the impact of the COVID-19 pandemic and measures taken in response to the pandemic; and changes in our operating plan.

These and other important factors discussed in our filings with the U.S. Securities and Exchange Commission (the SEC), including under the caption "Risk Factors" contained therein, could cause actual results to differ materially from those indicated by the forward-looking statements made in this press release. Any such forward-looking statements represent management's estimates as of the date of this press release. Except as required by law, we disclaim any intention or obligation to update or revise any forward-looking statements. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to the date of this press release.

Every Second Counts!®

Kiniksa Investor and Media Contact
Rachel Frank
(339) 970-9437
rfrank@kiniksa.com

Appendix 18.6.1(d)

Baseball Arbitration

[***]

[***] Certain information in this document has been excluded pursuant to Regulation S-K, Item 601(b)(10)(iv). Such excluded information is not material and would likely cause competitive harm to the registrant if publicly disclosed.

AMENDMENT NO. 2 TO ASSET PURCHASE AGREEMENT

This Amendment No. 2 to Asset Purchase Agreement (this "***Amendment No. 2***") is dated as of August 2, 2022 (the "***Amendment No. 2 Effective Date***") and is made by and between Biogen MA Inc., a Massachusetts corporation ("***Biogen***"), and Kiniksa Pharmaceuticals, Ltd., a Bermuda exempted company ("***Kiniksa***"). Kiniksa and Biogen are sometimes referred to herein individually as a "***Party***" and collectively as the "***Parties***." Terms used in this Amendment No. 2 and not otherwise defined shall have the respective meanings set forth in the APA (as defined below).

WHEREAS, pursuant to the terms of the Asset Purchase Agreement, dated as of September 7, 2016 (the "***APA Effective Date***"), by and between Biogen and Kiniksa, as amended by that certain Amendment No. 1 to Asset Purchase Agreement, dated July 31, 2017, by and between Biogen and Kiniksa (such agreement, as so amended, the "***APA***") Biogen agreed to sell to Kiniksa, and Kiniksa agreed to purchase from Biogen, certain assets of Biogen used in or relating to BIIB069 and BIIB22G11, all upon the terms and conditions set forth therein;

WHEREAS, simultaneously with the execution of this Amendment No. 2, Kiniksa Pharmaceuticals (UK), Ltd., a company incorporated under the laws of England and Wales ("***Kiniksa UK***") is entering into an agreement with Genentech, Inc., a Delaware corporation ("***Genentech***") and F. Hoffman-La Roche Ltd., a company under the laws of Switzerland ("***Roche***"; and together with Genentech, "***Licensee***" and such agreement the "***Licensee Agreement***"), pursuant to which Kiniksa UK will, effective as of the effective date of the Licensee Agreement (the "***Licensee Agreement Effective Date***"), grant to Licensee, among other things, a license or sublicense, as the case may be, under certain of the rights and licenses or sublicenses granted or otherwise transferred by Biogen to Kiniksa under the APA;

WHEREAS, simultaneously with the execution of this Amendment No. 2, Kiniksa and Kiniksa UK are executing an Asset Transfer Agreement pursuant to which, as permitted under Section 10.6 of the APA, Kiniksa has assigned to Kiniksa UK, and Kiniksa UK has assumed, in each case, all rights and obligations of Kiniksa under the APA, which includes this Amendment No. 2, effective as of the Licensee Agreement Effective Date;

WHEREAS, the Parties now wish to amend the APA and otherwise agree upon certain representations, warranties, and covenants in connection with the Licensee Agreement, in each case, to be effective solely as of the Licensee Agreement Effective Date (and which amendments and other representations, warranties, and covenants will *not* be effective prior to the Licensee Agreement Effective Date); and

WHEREAS, pursuant to Section 10.1 of the APA, no amendment, supplement or other modification to any provision of the APA shall be binding unless in writing and signed by both Parties.

NOW, THEREFORE, in consideration of the mutual covenants contained herein, and for other good and valuable consideration, the receipt and adequacy of which are hereby acknowledged, the Parties hereto, intending to be legally bound, hereby agree as follows:

1. Amendment to APA. Commencing on the Licensee Agreement Effective Date and thereafter for so long as the Licensee Agreement remains in effect, to the extent under the Licensee Agreement Kiniksa granted or provided to Licensee rights, licenses or other sublicenses with respect to Licensed Antibodies and Licensed Products (as each such term is defined in the Licensee Agreement) under the rights, licenses or sublicenses granted or otherwise transferred to Kiniksa from Biogen under the Biogen APA ("***Sublicensed Rights***"), the terms of the APA are hereby modified as set forth in Appendix A attached hereto solely with respect to the Sublicensed Rights. Notwithstanding any provision to the contrary set forth in this Amendment No. 2, upon termination or expiration of the Licensee Agreement in its entirety, the modifications to the terms of the APA set forth in this Section 1 and in Appendix A will terminate and all terms of the APA modified hereby with respect to such Sublicensed Rights will revert to the version of such terms in effect as of immediately prior to the Amendment No. 2 Effective Date.

2. Notice of Licensee Agreement Effective Date. Kiniksa shall promptly notify Biogen in writing of the occurrence of the Licensee Agreement Effective Date.

3. Miscellaneous. The Parties hereby confirm and agree that, except as amended hereby, the APA remains in full force and effect and continues to be a binding obligation of the Parties. This Amendment No. 2 may be executed in any number of counterparts, each of which will be deemed an original, but all of which together will constitute one and the same instrument. For purposes hereof, an e-Signature or email with attached pdf copy of this Amendment No. 2, including the signature pages hereto, will be deemed to be an original. The Parties agree that execution of this Amendment No. 2 by e-Signatures (as defined below) shall have the same legal force and effect as the exchange of original signatures. For the purposes of this Amendment No. 2, "***e-Signature***" shall mean a signature that consists of one or more letters, characters, numbers or other symbols in digital form incorporated in, attached to or associated with the electronic document, (a) that is unique to the person executing the signature; (b) the technology or process used to make the signature is under the sole control of the person making the signature; (c) the technology or process can be used to identify the person using the technology or process; and (d) the electronic signature can be linked with an electronic document in such a way that it can be used to determine whether the electronic document has been changed since the electronic signature was incorporated in, attached to or associated with the electronic document.

[**Remainder of page intentionally left blank**]

IN WITNESS WHEREOF, the Parties hereto have executed this Amendment No. 2 by their duly authorized representatives as of the Amendment No. 2 Effective Date.

KINIKSA PHARMACEUTICALS, LTD.

By: /s/ Mark Ragosa
Name: Mark Ragosa
Title: Senior Vice President & CFO

BIOGEN MA INC.

By: /s/ Michael Shih
Name: Michael Shih
Title: Head of Business Development

APPENDIX A

Capitalized terms not otherwise defined in this Appendix shall have the meaning given such terms in Amendment No. 2 to which this Appendix is attached and for purposes of interpreting this Appendix A as part of the APA, terms so defined in this Appendix A and Amendment No. 2 shall apply to the APA *mutatis mutandis*):

1. Net Sales Calculations.

The term "Net Sales" as used in the Biogen APA shall be deleted in its entirety and replaced with the following:

"***Net Sales***" means, for a Product in a particular period, (a) the amount calculated by subtracting from the Sales (as defined below) of such Product for such period: (i) a lump sum deduction of [***] of Sales in lieu of those deductions that are not accounted for on a Product-by-Product basis (e.g., freight, postage charges, transportation insurance, packing materials for dispatch of goods, custom duties); (ii) uncollectible amounts accrued during such period based on a proportional allocation of the total bad debts accrued during such period and not already taken as a gross-to-net deduction in accordance with the then currently used IFRS (as defined below) in the calculation of Sales of such Product for such period; (iii) credit card processing fees accrued during such period on such Sales and not already taken as a gross-to-net deduction in accordance with the then currently used IFRS in the calculation of Sales of such Product for such period; and (iv) government mandated fees and taxes and other government charges accrued during such period not already taken as a gross-to-net deduction in accordance with the then currently used IFRS in the calculation of Sales of such Product for such period, including, for example, any fees, taxes or other charges that become due in connection with any healthcare reform, change in government pricing or discounting schemes, or other action of a government or regulatory body; and (b) any Compulsory Sublicensee Compensation received by the GNE Group in such period. To the extent any of the deduction classes specified in clause (a) above are costs applicable to more than one GNE product, GNE shall allocate such costs among such products in a manner consistent with internal cost accounting practices. For clarity, no deductions taken in calculating Sales as specified below may be taken a second time in calculating Net Sales.

And for the purpose of calculating such Net Sales, ***"Sales"*** means for a Product in a particular period, the sum of (i) and (ii):

(i) the amount stated in the Roche Holding AG "Sales" line of its externally published audited consolidated financial statements with respect to such Product for such period (excluding sales to any sublicensees of Licensee that are not Affiliates (as defined in the Licensee Agreement) of Licensee. This amount reflects the gross invoice price at which such Product was sold or otherwise disposed of (other than for use as clinical supplies or free samples) by Licensee and its Affiliates to such third parties (excluding sales to any sublicensees that are not Affiliates of Licensee) in such period reduced by gross-to-net deductions, if not previously deducted from such invoiced amount, taken in accordance with the then currently used IFRS.

By way of example, the gross-to-net deductions taken in accordance with IFRS as of the Effective Date include items such as the following: [***].

For purposes of clarity, sales by Licensee and its Affiliates to any sublicensee shall be excluded from "Sales."

(ii) for sublicensees of Licensee that are not Licensee's Affiliates (and excluding Compulsory Sublicensees (as defined below)), the sales amounts reported to Licensee and its Affiliates in accordance with the sublicensee contractual terms and their then-currently used accounting standards. Notwithstanding anything to the contrary in the foregoing, Compulsory Sublicense Compensation shall be excluded from the "sales" amount.

Net Sales for Combination Products. Notwithstanding the foregoing, in the event a Product is sold as a Combination Product in any country in the Territory in any Calendar Quarter, the Net Sales on which royalties with respect to such Combination Product are payable shall be calculated by [***].

2. Additional Royalties for Net Sales of Product under the Licensee Agreement.

Kiniksa shall pay an additional [***] in addition to the royalty rate specified for each of the royalty tiers set forth in Section 5.3.1 of the Biogen APA. For clarity, Net Sales shall be calculated as set forth in this Appendix A and the calculation of royalties and respective royalty tiers under Section 5.3 of the Biogen APA shall be based upon the aggregate Calendar Year Net Sales by Licensee (and its Affiliates and sublicensees) under the Licensee Agreement of a given Product in the Territory during the applicable Royalty Term in the applicable country.

3. Adjustments to Royalties for Competing Drugs. For purposes of this Appendix A, Section 5.3.2(c) of the Biogen APA, shall be deleted in its entirety and replaced with the following:

Upon the first entry in a given country of a Competing Drug, the royalties in such country for such Product shall be reduced as follows: (a) If in any Calendar Quarter at any time after entry of a Competing Drug there has been a decline of the Sales of the applicable Product in such country greater than [***] of the level of the Sales of such Product achieved in [***] immediately prior to such entry, then the royalty payments due to Biogen for such Product in such country shall be reduced by [***]; and (b) if in any Calendar Quarter at any time after entry of a Competing Drug there has been a decline of the sales of the applicable Product in such country greater than [***] of the level of the sales of such Product achieved in [***] immediately prior to such entry, then the royalty payments due to Biogen for such Product in such country shall be reduced by [***]. For purposes of this Section 5.3.2(c), (x) a ***"Competing Drug"*** means, with respect to a Product, a therapeutic product that is not produced, licensed or owned by Licensee, its Affiliates or sublicensees and that (i) [***], (ii) [***] and (iii) [***]; and (y) "Bioequivalent" or "Bioequivalence" means, a biological product that (i) is highly similar to the Product notwithstanding minor differences in clinically inactive components; and (ii) has no clinically meaningful differences between the biological product and the Product in terms of the safety, purity, and potency.

4. Sales Milestones Accounting Period Calculations. The calculations for whether Net Sales for a given Product has achieved the respective sales threshold milestone event tiers as set forth in Section 5.2.3 of the Biogen APA will be calculated [***].

5. Indication. The term "Indication" as used in the Biogen APA shall be deleted in its entirety and replaced with the following:

"***Indication***" means any human indication, disease or condition in the Field that can be treated, prevented, cured or the progression of which can be delayed, excluding an expansion of label claim for an already approved indication. Examples of label expansion include [***].

6. Product. The term "Product" as used in the Biogen APA shall be deleted in its entirety and replaced with the following:

"***Product***" means any product, including any Combination Product, that (a) contains or incorporates any Acquired Antibody or (b) the manufacture, use or sale of which is Covered by a Valid Claim. [***].

7. Combination Product. The term "Combination Product" as used in the Biogen APA shall be deleted in its entirety and replaced with the following:

"***Combination Product***" means a Product that includes (a) a single pharmaceutical formulation containing as its active ingredients both an Acquired Antibody and one or more other therapeutically or prophylactically active ingredients; (b) a combination therapy comprised of an Acquired Antibody and one or more other therapeutically or prophylactically active products, priced and sold in a single package containing such multiple products or packaged separately but sold together for a single price; or (c) a combination therapy comprised of an Acquired Antibody and a Companion Diagnostic, priced and sold in a single package containing such multiple products or packaged separately but sold together for a single price; in each case (a) through (c), including all dosage forms, formulations, presentations, line extensions, and package configurations. Each of the therapeutically or prophylactically active ingredients or products, other than the Acquired Antibody, referred to in the foregoing clause (a) or (b) is an "***Other Component***."

8. Valid Claim. The term "Valid Claim" as used in the Biogen APA shall be deleted in its entirety and replaced with the following:

"***Valid Claim***" means (a) an issued and unexpired claim within the Acquired Patent Rights, the Kiniksa Patent Rights, the Background Licensed Patent Rights or the Background Sublicensed Intellectual Property that has not been disclaimed, revoked or held invalid by a final non-appealable decision of a court of competent jurisdiction or government agency or (b) a claim of a pending patent application within the Acquired Patent Rights, the Kiniksa Patent Rights, the Background Licensed Patent Rights or the Background Sublicensed Intellectual Property that, in the case of any such patent application, was filed in good faith, has not been pending for more than [***], and has not been abandoned or finally disallowed.

9. Additional Supporting Definitions. For the purposes of this Appendix A and application of such terms in the Biogen APA the following terms, whether used in the singular or plural, shall have the following meanings:

"***Companion Diagnostic***" means any product that is used for predicting or monitoring the response of a human being to treatment with a Product (e.g., device, compound, kit, biomarker or service that contains a component that is used to detect or quantify the presence or amount of an analyte in body or tissue that affects the pathogens of the disease).

"***Compulsory Sublicense***" means, for a given country or region, a license or sublicense of patent rights licensed (or sublicensed) by Kiniksa to Licensee granted to a third party (a "***Compulsory Sublicensee***") through the order, decree or grant of a governmental authority having competent jurisdiction in such country or region, authorizing such third party to manufacture, use, sell, offer for sale, import or export a Product in such country or region.

"Compulsory Sublicense Compensation" means for a given country or region in the Territory, the compensation paid to GNE by a Compulsory Sublicensee for its Compulsory Sublicense.

"***IFRS***" means International Financial Reporting Standards.

"***regulatory exclusivity***", whether or not capitalized, shall mean, with respect to any Product in any country or jurisdiction in the Territory, the period of time during which (a) a Party or its Affiliate or sublicensee has been granted any exclusive marketing rights, other than a Patent Right, by any governmental authority under applicable law that prevents Third Parties from selling such Product in such country or jurisdiction, including orphan drug exclusivity, pediatric exclusivity, rights conferred in the U.S. under the FD&C Act, in the European Union under Directive 2001/83/EC, or rights similar thereto in other countries or regulatory jurisdictions in the Territory; or (b) the data and information submitted by a Party or its Affiliate or sublicensee to the relevant Regulatory Authority in such country or jurisdiction for purposes of obtaining Marketing Authorization of such Product may not be disclosed, referenced, or relied upon in any way by any Third Party or such Regulatory Authority to support the marketing authorization of any product by any Third Party in such country or jurisdiction, or if such data and information is disclosed, referenced, or relied upon to support a marketing authorization for such product granted to any Third Party in such country or jurisdiction, then such product may not be placed on the market for any indication.

FIFTH AMENDMENT OF SUBLEASE

FIFTH AMENDMENT OF SUBLEASE (the "Fifth Sublease Amendment") made as of the 27th day of July, 2022, by and between 92 HAYDEN AVENUE TRUST, as landlord ("Landlord"), and KINIKSA PHARMACEUTICALS CORP., a Delaware corporation, as tenant (interchangeably called "Kiniksa" or "Tenant").

RECITALS

WHEREAS, by Lease dated May 22, 2008, as amended by First Amendment to Lease dated November 24, 2015 (collectively, the "Lease"), Landlord did lease to Shire Human Genetic Therapies, Inc., as successor-in-interest to AMAG Pharmaceuticals, Inc. ("Original Tenant") and Original Tenant did lease and hire from Landlord the entirety of the building on the site known as 100 Hayden Avenue, Lexington, Massachusetts (the "Building") as more particularly described in the Lease (the "Premises");

WHEREAS, by Sublease Agreement dated as of March 18, 2018 between Original Tenant, as sublandlord, and Kiniksa, as subtenant (the Sublease Agreement as amended by and affected by the documents listed on Exhibit A attached hereto is hereinafter referred to as the "Sublease"), subleases the entirety of the Premises (referred to in the Sublease as the "Subleased Premises" and hereinafter as either the Premises or the Subleased Premises) upon the terms set forth in the Sublease;

WHEREAS, by Recognition and Attornment Agreement and Amendment of Sublease dated as of November 6, 2020 between Landlord and Kiniksa (the "Fourth Sublease Amendment"), (i) Kiniksa leased the Premises directly from Landlord upon the expiration of the Lease and Landlord recognized the Sublease as a direct lease between Landlord and Kiniksa and (ii) Landlord and Kiniksa extended the Term of the Sublease, upon the terms set forth in such Fourth Sublease Amendment;

WHEREAS, the Term of the Sublease is scheduled to expire on August 31, 2023 (the “Sublease Expiration Date”) and Landlord and Kiniksa have agreed to further extend the term of the Sublease upon all the same terms and conditions contained in the Sublease, except only as otherwise specifically modified by this Fifth Sublease Amendment; and

WHEREAS, Landlord and Kiniksa are entering into this Fifth Sublease Amendment to set forth said agreement.

NOW, THEREFORE, in consideration of One Dollar ($1.00) and other good and valuable consideration paid by each of the parties to the other, the receipt and sufficiency of which is hereby acknowledged, and in further consideration of the provisions herein contained, Landlord and Kiniksa do hereby covenant and agree as follows:

1. The term of the Sublease, which but for this Fifth Sublease Amendment is scheduled to expire on August 31, 2023, is hereby extended for the period commencing on September 1, 2023 and expiring on August 31, 2024 (the "Second Extended Term"), unless sooner terminated in accordance with the provisions of the Sublease, upon all of the same terms and conditions contained in the Sublease *mutatis mutandis*, except as specifically amended by this Fifth Sublease Amendment (the Sublease, as modified and extended by this Fifth Sublease Amendment is herein referred to as the "Amended Sublease").

2. Monthly Base Rent shall be payable by Kiniksa to Landlord from September 1, 2023 through August 31, 2024 in the monthly amount of $247,207.38.

3. For the sake of clarity, Kiniksa shall have no further option to extend the Term of the Sublease beyond the Second Extended Term.

4. (a) Tenant warrants and represents that Tenant has not dealt with any broker in connection with the consummation of this Fifth Sublease Amendment other than Colliers International (the "Broker") and in the event any claim is made against Landlord relative to dealings by Tenant with brokers other than the Broker, Tenant shall defend the claim against Landlord with counsel of Tenant's selection first approved by Landlord (which approval will not be unreasonably withheld or delayed) and save harmless and indemnify Landlord on account of loss, cost or damage which may arise by reason of such claim.

5. (b) Landlord warrants and represents that Landlord has not dealt with any broker in connection with the consummation of this Fifth Sublease Amendment other than the Broker; and in the event any claim is made against Tenant relative to dealings by Landlord with brokers, Landlord shall defend the claim against Tenant with counsel of Landlord's selection and save harmless and indemnify Tenant on account of loss, cost or damage which may arise by reason of such claim. Landlord shall be responsible for the payment of the brokerage commission to the Broker respecting the Second Extended Term pursuant to a separate agreement between the parties.

6. Each of Landlord and Kiniksa acknowledges, covenants and agrees that, as of the date of this Fifth Sublease Amendment, it has no demands, causes of action, claims or other actions against the other under the Sublease.

7. This Fifth Sublease Amendment shall be binding on and shall inure to the benefit of the Landlord and Kiniksa and their respective successors and assigns as of the date of this Fifth Sublease Amendment.

8. Except as only as specifically amended herein, the Sublease shall remain unchanged and in full force and effect.

9. The parties acknowledge and agree that this Fifth Sublease Amendment may be executed by electronic signature, which shall be considered as an original signature for all purposes and shall have the same force and effect as an original signature. Without limitation, "electronic signature" shall include faxed versions of an original signature, electronically scanned and transmitted versions (e.g., via pdf) of an original signature or signatures transmitted through any electronic method complying with the federal ESIGN Act (e.g., DocuSign).

[Remainder of page intentionally left blank]

EXECUTED as of this date and year first above written.

92 HAYDEN AVENUE TRUST

/s/ Patrick M. Mulvihill

Patrick M. Mulvihill, For the Trustees of 92 Hayden Avenue Trust, Pursuant to Written Delegation, but not individually

KINIKSA PHARMACEUTICALS CORP.

By: /s/ Mark Ragosa

Name: Mark Ragosa

Title: Senior Vice President, Chief Financial Officer

By: /s/ Madelyn Zeylikman

Name: Madelyn Zeylikman

Title: General Counsel Senior Vice President and Secretary

EXHIBIT A

SUBLEASE DOCUMENTS

1. Sublease Agreement dated March 13, 2018 between Original Tenant and Kiniksa
2. Consent to Sublease dated March 13, 2018 among Landlord, Original Tenant and Kiniksa
3. First Amendment to Sublease Agreement dated June 26, 2018 between Original Tenant and Kiniksa
4. Letter Agreement consenting to First Amendment to Sublease dated June 25, 2018 among Landlord, Original Tenant and Kiniksa
5. Second Amendment to Sublease Agreement dated July 17, 2018 between Original Tenant and Kiniksa
6. Third Amendment to Sublease Agreement dated November 7, 2018 between Original Tenant and Kiniksa
7. Letter Agreement consenting to Second Amendment to Sublease and Third Amendment to Sublease dated November 8, 2018 among Landlord, Original Tenant and Kiniksa
8. Letter Agreement dated August 2, 2019 among Landlord, Original Tenant and Kiniksa (the "Analytical Lab Consent")
9. Fourth Amendment to Sublease Agreement dated November 6, 2020 between Original Tenant and Kiniksa (consented to by Landlord in the Agreement to which this exhibit is attached)

Exhibit 31.1

CERTIFICATIONS

I, Sanj K. Patel, certify that:

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

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

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

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

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

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

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

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

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

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

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

November 3, 2022

/s/ Sanj K. Patel

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

Exhibit 31.2

CERTIFICATIONS

I, Mark Ragosa, certify that:

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

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

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

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

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

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

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

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

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

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

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

November 3, 2022

/s/ Mark Ragosa

Mark Ragosa
Chief Financial Officer
(Principal Financial Officer)

Exhibit 32.1

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

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

(1) The Quarterly Report on Form 10-Q of the Company for the period ended September 30, 2022 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and

(2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

November 3, 2022

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

Exhibit 32.2

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

I, Mark Ragosa, 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 September 30, 2022 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and

(2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

November 3, 2022

/s/ Mark Ragosa
Mark Ragosa
Chief Financial Officer
(Principal Financial Officer)